Solid Tumor Rules

Effective with Cases Diagnosed 1/1/2018 and Forward

2024 Update

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Solid Tumor Rules
Preface
(Excludes lymphoma and leukemia M9590 – M9993)

In Appreciation

NCI SEER gratefully acknowledges the dedicated work of Dr. Charles Platz who has been with the project since the inception of the 2007 Multiple Primary and Histology Coding Rules. We appreciate the support he continues to provide for the Solid Tumor Rules. The quality of the Solid Tumor Rules directly relates to his commitment.

NCI SEER would also like to acknowledge the Solid Tumor Work Group who provided input on the manual. Their contributions are greatly appreciated.

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The Solid Tumor Rules

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Preface

The 2007 Multiple Primary and Histology (MPH) Coding Rules have been revised and are now referred to as *the Solid Tumor Rules.* The 2007 Multiple Primary and Histology Coding Rules were developed to promote consistent and standardized coding by cancer registrars and the Solid Tumor Rules continue to provide coding instructions to ensure accurate data collection. It is important to note that eight site specific coding modules were updated for 2018. These site groups are: Benign Brain, Malignant CNS, Breast, Colon, Lung, Head & Neck, Kidney, and Urinary. Cutaneous melanoma was updated for 2021, and Other Sites was updated for 2023 (the 2007 MPH Other Sites applies to Other Sites cases diagnosed 2007-2022).

The primary reference for both the 2007 MPH rules and Solid Tumor Rules are the WHO Classification of Tumors books (blue books). Since 2007, WHO has continued publishing updates to the WHO Classification of Tumors series. As part of each new edition, subject matter experts review current literature and make recommendations regarding current practices in histology terminology and diagnosis. The College of American Pathologists (CAP) has adopted the new histologic terminology and diagnosis criteria into the site-specific 2018 CAP Protocols. The Solid Tumors Rules have been revised to reflect current CAP and WHO practices.

As part of the revisions to the 2007 MPH rules, the editors and Solid Tumor Committee reviewed issues and questions NCI SEER received since the implementation of the MPH rules. These questions provided valuable information as to what clarifications were needed in the form of additional rules, tables, examples, and notes.

The Solid Tumor Committee has members who represent all the standard setters including NCI SEER, American College of Surgeons (ACoS), Commission on Cancer (CoC), American Joint Commission on Cancer (AJCC), the Centers for Disease Control and Prevention (CDC), National Program of Cancer Registries (NPCR), North American Association of Central Cancer Registries (NAACCR), the Canadian Cancer Registry (CCR), and central and hospital cancer registries.

Physician guidance by specialty pathologists and clinicians was integral to the review and revision process. Regular consultation with the editors of the WHO Classification of Tumors series and AJCC physicians ensured that the new rules accurately reflect the editors' intent and purpose.

Solid Tumor Rules
General Instructions
(Excludes lymphoma and leukemia M9590 – M9993)

What You Need to Know About the Solid Tumor Rules

There are specific instructions preceding each set of histology rules that define terms which may be used to code histology as well as terms which may not be used to code histology. These changes are in accordance with current WHO and CAP guidelines.

Eight site groups were revised for 2018. The Solid Tumor General Instructions apply to the revised sites listed below:

- Head & Neck
- Colon (includes rectosigmoid and rectum for cases diagnosed 1/1/2018 forward)
- Lung
- Breast
- Kidney
- Urinary sites
- Non-malignant CNS
- Malignant CNS and Peripheral Nerves

Cutaneous Melanoma was updated for 2021. The Solid Tumor General Instructions apply to cutaneous melanoma cases diagnosed January 1, 2021 and forward.

The 2007 Multiple Primary & Histology rules and the 2007 General Instructions are to be used for cases diagnosed 1/1/2007 to 12/31/2022 for the following site group:

- Other Sites
    - Primary sites excluded are:
        - Rectosigmoid and rectum which are included in the Solid Tumor Colon rules
        - Peripheral nerves which are included in the Solid Tumor Malignant Brain and Head & Neck rules

Other Sites was updated for 2023. The Solid Tumor General Instructions apply to sites covered in Other for cases diagnosed January 1, 2023 and forward.

Solid Tumor Rules
General Instructions
(Excludes lymphoma and leukemia M9590 – M9993)

SUBMITTING QUESTIONS

Submit technical questions and suggestions related to this manual to Ask a SEER Registrar on the SEER website. SEER regions may also submit technical questions to NCI SEER inquiry system using the web-based SINQ system. When submitting questions, make sure you select the correct category (2007 MPH rules or Solid Tumor Rules) AND **always include primary site and diagnosis year.**
IMPORTANT INFORMATION: When needed, we will consult with experts to provide guidance and clarifications when answering difficult or unusual questions. Our specialty matter experts (SMEs) are authors of WHO Classification of Tumors books, CAP pathologists, and recognized experts in their fields of interest.

General Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Adenocarcinoma; glandular carcinoma; carcinoma
- De novo; new tumor; frank (obsolete term)
- Multicentric; multifocal
- No evidence of disease; NED; disease free
- Simultaneous; synchronous; at the same time; prior to first course treatment
- Topography; site code
- Tumor; mass; tumor mass; lesion; neoplasm; nodule
  - The terms tumor, mass, tumor mass, lesion, neoplasm and nodule are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician's statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to determine multiple primaries
  - **Do not** use these terms for casefinding or determining reportability
- Type; subtype; variant

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How to Navigate the Solid Tumor Rule Modules

The PDFs must be opened in Adobe Reader for complete functionality of content controls. If the PDF document opens in your browser by default, contact your IT department to change the settings for your browser.

The following functions will help you maneuver within site groups.

1. **Navigating between hyperlinks:** When you use a hyperlink to go to another place in the rules, use the PREVIOUS VIEW button to return to your starting point. For example, a hyperlink in the Equivalent Terms and Definitions sends you to the **Histology Rules.** When you are finished with the histology rules, click the PREVIOUS VIEW button to return to the location of the hyperlink in the Equivalent Terms and Definitions.
   Note 1: **To enable this button**, right click on the toolbar, select Page Navigation and click Previous View. A left-pointing arrow will appear on the toolbar.
   Note 2: If you scroll through multiple pages after using a hyperlink, the Previous View button returns to the most recently viewed page. This means that you must click the Previous View button **multiple times** to return to your starting point.
2. **Bookmarks:** In the left panel, use the bookmarks to quickly jump between sections and subsections.
   A. Click the [+] to expand a bookmark level, showing all of the sub-levels.
   B. Click the [-] to collapse a bookmark level, showing a main level.
3. **Footer links:** Alternatively, there are links in the footer of every page that go to the first page of other sections within a site group.
4. **The Search Function:** Pressing CTRL + F will display a search box. Enter the desired term in the search box and press ENTER or NEXT. When there are multiple occurrences of the term:
   A. Use the NEXT button to view consecutive occurrences of the term.
   B. Use the PREVIOUS button go back to the most recently viewed term.
5. **Adobe Reader/Acrobat Updates:** Be sure to keep your version of Adobe up to date. The above functions may not work on outdated versions. You may need to consult your IT department to obtain the latest versions.
6. For full performance on mobile operating systems, it is recommended that you download the free Adobe Reader app from the Apple App Store or Google Play Store.

Solid Tumor Rules
General Instructions
(Excludes lymphoma and leukemia M9590 – M9993)

How to Use the Solid Tumor Rules

Note: The rules do not apply to hematopoietic primaries (lymphoma and leukemia) of any site. Use the Hematopoietic & Lymphoid Neoplasm Coding Manual and Database for histologies M9590-M9993.

1. The **purpose** of these rules is to determine **multiple primaries** and to code **histology ONLY**. The Solid Tumor Rules are **not used** to determine case reportability, casefinding, stage, or tumor grade. For instructions on coding grade, stage, SSDIs, and treatment, please refer to the appropriate manuals.
2. Staging systems are **not used** to determine the number of primaries or histology.
3. Use the following site-specific rules for tumors diagnosed 1/1/2018 and forward:
    - Malignant CNS and Peripheral Nerves
    - Non-Malignant CNS
    - Breast
    - Colon
    - Head and neck
    - Kidney
    - Lung
    - Urinary sites
4. Use the following site-specific rules for tumors diagnosed 1/1/2021 forward:
    - Cutaneous Melanoma
5. Use the following site-specific rules for tumors diagnosed 1/1/2007 through 12/31/2022:
    - 2007 MPH Other Sites (not updated for 2018) for solid tumors which occur in primary sites not covered by the site-specific rules.
6. The appropriate rule set to use is based on **date of diagnosis**.
    - Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules
    - Tumors diagnosed 01/01/2018 and later: Use the Solid Tumor Rules (with exceptions in #4 and 5)
    - An original tumor diagnosed before 1/1/2018 and a subsequent tumor diagnosed 1/1/2018 or later **in the same primary site**: Use the Solid Tumor Rules
    - A melanoma diagnosed before 1/1/2021 and a subsequent melanoma diagnosed 1/1/2021 or later: Use the 2021 Cutaneous Melanoma Rules
    - A primary using Other Sites MPH rules diagnosed before 1/1/2023 and a subsequent primary also covered in Other Sites diagnosed 1/1/2023 or later: Use the 2023 Other Sites Rules
7. Use the Solid Tumor Rules in the following order:
    A. For multiple tumors, you must decide whether they are a single or multiple primaries:
        i. Use the Histology Rules to assign a "working" histology for each tumor.

ii. Use Multiple Primary Rules to determine whether the tumors are a single primary or multiple primaries.
iii. If a single primary, follow the priority order in #7B.
iv. If multiple primaries, follow the priority order in #7B for **EACH** of the separate tumors/primaries.

B. For a single tumor or multiple tumors determined to be a single primary:
i. General Instructions
ii. Equivalent Terms and Definitions
iii. Multiple Primary Rules
iv. Histology Rules

8. The Solid Tumor Rules are available in text format.
9. **Notes** and **examples** are included with some of the rules to highlight key points or to add clarity to the rules.
10. Rules are in **hierarchical** order within each module. Use the first rule that applies and

STOP

How to Use the Equivalent Terms and Definitions

The Equivalent Terms and Definitions contain the following:

- Changes from the 2007 Multiple Primary and Histology Rules
- Equivalent and equal terms
- Terms that are not equivalent or equal
- Tables for coding
  - Primary site codes
  - Combination histologies
  - Reportable histologies and subtypes/variants
  - Not reportable histologies
  - Paired sites
- Illustrations

Solid Tumor Rules
General Instructions
(Excludes lymphoma and leukemia M9590 – M9993)

Multiple Primary Rules Do NOT Apply to Tumors Described as Metastases

Each Multiple Primary Rule section begins with a note that reads, "These rules are **NOT** used for tumor(s) described as metastases." This means that a tumor in a **metastatic site** is **not counted** when deciding which module to use in the Multiple Primary Rules (**Unknown if Single or Multiple Tumors**, **Single Tumor or Multiple Tumors**).

Example: Tumor located in the left lung diagnosed as metastatic breast cancer. Do not use the Lung Solid Tumor Rules to determine if this is a new primary. If the patient is known to have a breast primary, the lung tumor is recorded as metastasis from that primary. If the patient is unknown to have a breast primary, then refer to the Breast Solid Tumor Rules as the lung findings are stated to originate in the breast.

How to Use the Multiple Primary Rules

1. To choose the appropriate module (Unknown if Single or Multiple Tumors, Single Tumor, Multiple Tumors), determine the **number of tumors**.
   A. Do not count **metastatic** lesions when determining which module to use.
   B. When the number of tumors is **unknown/not documented**, use the "Unknown if Single or Multiple Tumors" module.
      i. When there is a tumor or tumors with separate microscopic foci, ignore the microscopic foci.
   C. When the patient has a **single tumor**, use the "Single Tumor" module.
   D. When the patient has **multiple tumors**, use the "Multiple Tumors" module.
2. When the rules return a single primary, prepare one abstract.
3. When the rules return multiple primaries, prepare two or more abstracts.
4. For those sites/histologies which have recognized **biomarkers**, the biomarkers frequently identify the histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.
5. Do not use physician staging to determine multiple primaries.

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Timing Rules

Each Solid Tumor site group includes timing rules in the Multiple Primary Rules. It is important to remember that timing rules differ by site. Examples of timing rules include:

- Abstract **multiple primaries** when the patient has a subsequent tumor after being **clinically disease-free** for **greater than X years** after the original diagnosis or last recurrence.
- Abstract a **single primary** (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60** days **after** an **in-situ** tumor

One year = 365 days
Example: A tumor diagnosed 1/1/2018 and second tumor diagnosed 1/1/2019 occur exactly one year apart.

Less than one year = Less than 365 days

More than one year = 366 days or more
Example: A tumor diagnosed 1/1/2018 and second tumor diagnosed 1/2/2019 occur more than one year apart.

- No evidence of disease (NED) means complete response to treatment.
- **Clinically disease-free** means that there was **no <u>evidence</u>** of recurrence on follow-up.
    - Once a patient has been diagnosed with **metastatic disease**, whether at diagnosis or later, they will never be NED.
- When there is a recurrence less than or equal to X years of diagnosis, the **"clock"** starts over. The time interval is calculated from the **date of last recurrence.** In other words, the patient must have been disease-free for greater than X years from the date of the last recurrence.
- When it is **unknown/not documented** whether the patient had a recurrence, default to **date of diagnosis** to compute the time interval.
- Use the Multiple Primary Rules as written to determine whether a subsequent tumor is a new primary or a recurrence. The **ONLY exception** is when a **pathologist compares slides** from the subsequent tumor to the "original" tumor and documents the subsequent tumor is a recurrence of the previous primary. Never code multiple primaries based only on a physician's statement of "recurrence" or "recurrent".

Solid Tumor Rules
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Histologic Type ICD-O-3

Item Length: 4
NAACCR Item #: 522
NAACCR Name: Histologic Type ICD-O-3

The data item Histologic Type ICD-O-3 describes the microscopic composition of cells and/or tissue for a specific primary. The tumor type or histology is a basis for staging and determination of treatment options. Histology affects treatment decisions, prognosis and course of the disease.

The *International Classification of Diseases for Oncology*, Third Edition (ICD-O-3) is the standard reference for histology codes for tumors diagnosed in 2001 and later. Do not record the 'M' that precedes the histology code. See sections *Coding Guidelines for Topography and Morphology* and *Summary of Principal Rules for Using the ICD-O*, Third Edition for guidance in using the ICD-O-3.

Important Information for Coding Histologic Type for Cases Diagnosed 1/1/2018 Forward

The North American Association of Central Registries (NAACCR) release updates that contain new ICD-O codes, changes in behaviors for existing ICD-O codes, and new preferred terminology. The updates are, in chronological order:

- 2018 Guidelines for ICD-O-3 Histology Code and Behavior Update effective for cases diagnosed 1/1/2018 forward
- 2021 Guidelines for ICD-O-3 Histology Code and Behavior Update, effective for cases diagnosed 1/1/2021 forward
- 2022 Guidelines for ICD-O-3.2 Histology Code and Behavior Update, effective for cases diagnosed 1/1/2022 forward
- 2023 Guidelines for ICD-O-3.2 Histology Code and Behavior Update, effective for cases diagnosed 1/1/2023 forward

The Solid Tumor Editors recommend coding histology using:

- The Solid Tumor Rules
- The 2021 Cutaneous Melanoma Solid Tumor Rules
- Updated ICD-O histology codes and terms
- The ICD-O-3.2

When a histology code cannot be identified using the above recommendations, submit a question to Ask a SEER Registrar.

Solid Tumor Rules
General Instructions
(Excludes lymphoma and leukemia M9590 – M9993)

How to Use the Histology Rules

Note 1: **Do not use these rules to determine case reportability.**
Note 2: Refer to the How to Use the Solid Tumor Rules for instructions on the order in which to use the rules.

1. Rules are divided into two sections: Single Tumor and Multiple Tumors Abstracted as a Single Primary
   A. Each section is a complete set of rules.
   B. Within each section, the rules are hierarchical. Use the first rule that applies and **STOP**. Do not continue through the rules.
2. Code the histology diagnosis prior to **neoadjuvant therapy**. Neoadjuvant therapy can change the histological profile of the tumor. ***See site-specific modules for exceptions to this rule.***
3. Code the histology assigned by the physician. **Do not change histology** in order to make the case applicable for **staging**.
4. A list of terms which can be used and terms which cannot be used to code histology precede each set of histology rules.
5. Code a histology when described by ambiguous terminology **ONLY** when:
   - Histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.)
   - Patient is treated for the histology described by an ambiguous term
   - Case is accessioned (added to your database) based on a single histology described by ambiguous terminology and no other histology information is available/documented

Note: If the histology described by ambiguous terminology does not meet any of the criteria in bullets 1, 2, or 3, **DO NOT CODE** the histology.

Ambiguous Terminology

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

Ambiguous terminology from the SEER Manual and CoC Manual is used to determine reportability, not to determine histology.

Priority Order for Using Documentation to Code Histology

For each site, priorities include tissue/histology, cytology, radiography/scans, and physician diagnoses, and biomarkers. **You must use the priority order that precedes the histology rules for each site.**

- Priority order will differ by site. Tissue pathology (and/or biomarkers, if applicable) always takes precedence.
- The specific types of radiography/scans also differ by site.

Which document to use when there is conflicting information between the final diagnosis, synoptic report, or CAP protocol:

When there are discrepancies between the final diagnosis and synoptic report, use the document that provides the more specific histology. This will likely be found in the synoptic report. The CAP Protocol should be used only when a final diagnosis or synoptic report are not available. Definitions for CAP Protocol, final diagnosis, and synoptic report can be found in the Definitions section.

Definitions

***Note*: Use these terms and definitions for all reportable tumors except lymphoma and leukemia primaries (M9590-9993).**

Bilateral: Relating to the right **and** left sides of the body or of a body structure; bilaterality is **not** an indication of single or multiple primaries.

CAP Protocol: The CAP Cancer Reporting Protocols provide guidelines for collecting the essential data elements for complete reporting of malignant tumors and optimal patient care. The protocol is a check list which allows the pathologist to note their observations while reviewing the slides and/or gross specimen. CAP Protocols include all relative data elements including site, surgical procedure, tumor size, histology, grade, margins, lymph node status, and staging along with other site specific elements. The protocols are multiple pages.

Clinical Diagnosis: A diagnosis that is not microscopically confirmed. It may be based on information from the clinician's expertise.

Contiguous tumor: A single tumor that involves, invades, or bridges adjacent or connecting sites or subsites.

De novo: For colon cancer, de novo (formerly called frank) carcinoma originates in the mucosa of the colon rather than in a polyp.

Final Diagnosis: The final diagnosis is found in the pathology report. The findings from the CAP Protocol are consolidated into paragraph format.

Focal: An adjective meaning limited to one specific area. A focal cancer is limited to one specific area or organ. The area may be microscopic **or** macroscopic.

Foci: Plural of focus.

Focus: A term used by pathologists to describe a group of cells that can be **seen only by a microscope**. The cells are noticeably different from the surrounding tissue either by their appearance, chemical stain, or other testing.

Laterality: Indication of which side of a **paired organ/site** a tumor is located. (See Paired organ/site)

Solid Tumor Rules
General Instructions
(Excludes lymphoma and leukemia M9590 – M9993)

Multiple primaries: More than one reportable case.

NED: Acronym for "no evidence of disease"; disease free

Non-contiguous: Not touching along the boundary; not being in actual contact

Overlapping tumor: A single tumor which has spread from the primary site to adjacent organs or tissue. Adjacent organs/tissue are next to each other.

Paired organ/site: There are two organs, one on the left side of the body and one on the right side of the body. (See Laterality)

Recurrence: This term has two meanings:

- The reappearance of disease that was thought to be cured or inactive (in remission). Recurrent cancer starts from cancer cells that were not removed or destroyed by the original therapy.
- A new occurrence of cancer in the same primary site such as a previous adenocarcinoma of the right lung and a subsequent squamous cell carcinoma of the left lung called a "recurrence" of lung cancer (the patient had lung cancer before, now has another lung cancer). This type of recurrence arises from cells that have nothing to do with the earlier (first) cancer. A new or another occurrence, incidence, episode, or report of the same disease (cancer) in a general sense – a new occurrence of cancer.

Simultaneous: This term is used in the Solid Tumor Rules to describe malignant tumors diagnosed at the same time or during initial workup (prior to first course of therapy).

Single primary: One reportable case. The Multiple Primary Rules say "abstract a single primary" when multiple tumors are:

- Simultaneous and abstracted as a single primary **OR**
- Subsequent tumor(s) which are a recurrence rather than a multiple primary

Synchronous: See "Simultaneous".

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Synoptic Report: All core and conditionally required data elements outlined on the surgical case summary from the cancer protocol must be displayed in synoptic report format. Synoptic format is defined as:

- Data element: followed by its answer (response).
- The data element should be represented in the report as it is listed in the case summary. The response for any data element may be modified from those listed in the case summary, including "Cannot be determined" if appropriate.
- Each diagnostic parameter pair (Data element: Response) is listed on a separate line or in a tabular format to achieve visual separation. The following exceptions are allowed to be listed on one line:
    - Anatomic site or specimen, laterality, and procedure
    - Pathologic Stage Classification (pTNM) elements
    - Negative margins, as long as all negative margins are specifically enumerated where applicable
- The synoptic portion of the report can appear in the diagnosis section of the pathology report, at the end of the report or in a separate section, but all Data element: Responses must be listed together in one location

Unilateral: Relating to one side of the body or one side of a body structure

WHO/IARC: The International Agency for Research on Cancer (IARC) is a specialized cancer agency at the World Health Organization (WHO). WHO's primary role is to direct international health within the United Nations system and to lead partners in global health responses. IARC is responsible for the WHO Classification of Tumours, also known as the Blue Books.

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Introduction

Note 1: Breast includes Nipple C500; Central portion of breast C501; Upper-inner quadrant C502; Lower-inner quadrant C503; Upper-outer quadrant of breast C504; Lower-outer quadrant C505; Axillary tail C506; Overlapping lesion of breast C508; Breast NOS C509.

Note 2: Tables and rules refer to ICD-O rather than ICD-O-3. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

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

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

Note 4: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Biomarkers may identify the histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Changes from 2007 MPH Rules

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

1. **NST (N**o **S**pecial **T**ype**), mammary carcinoma NST, and carcinoma NST** are the new terms for duct or ductal carcinoma. Previously, it was thought that carcinoma originated in the ducts or lobules of the breast, hence the names duct carcinoma and lobular carcinoma. Current thinking is that carcinoma originates in the "terminal duct lobular unit" therefore the preferred term is NST or carcinoma NST.
2. **Mammary carcinoma** is a synonym for carcinoma no special type (NST)/duct carcinoma not otherwise specified (NOS) **8500.** It will **no longe**r be coded as carcinoma NOS **8010.**
3. **DCIS/Carcinoma NST in situ** has a major classification change.
   A. Subtypes/variant, architecture, pattern, and features **ARE NOT CODED**. The majority of in situ tumors will be coded to DCIS **8500/2.**
   B. It is very important to code the grade of all DCIS.
      i. Code grade as designated in current AJCC Manual, SEER Coding Manual, and COC Coding Manual.
      ii. The current breast **WHO** edition emphasizes coding the **grade** of tumor rather than the **subtype/variant**.
      iii. The WHO editions are used internationally by pathologists to keep their nomenclature and histology identification current.
      iv. Over time, **subtypes/variants** will be diagnosed **less frequently.**
4. The invasive subtype/variant is coded **ONLY** when it comprises **greater than 90%** of the tumor. This change has been implemented in both the WHO and in the CAP protocols.
5. **New codes/terms** are identified by asterisks (*) in the histology table in the Terms and Definitions.
6. Excerpt from the CAP Invasive Breast Protocol (page 17): "A modified list is presented in the protocol based on the most frequent types of invasive carcinomas and terminology that is in widespread usage. The modified list is intended to capture the majority of tumors and reduce the classification of tumors being reported as 'other.' The WHO classification is presented for completeness".

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

New for 2023

The rules for determining single versus multiple primaries in tumors with carcinoma NST/duct and lobular carcinoma have been revised and now align with ICD-O-3.2. Applicable Histology Rules have also been revised to reflect ICD-O-3.2 histology terminology and corresponding ICD-O codes.

New for 2024

1. Beginning with cases diagnosed 1/1/2024 forward, in situ lobular carcinoma with other types of in situ carcinoma 8524/2 has been deemed biologically impossible based on expert pathologist review for the Cancer PathCHART project. Table 2 has been updated with coding instructions and new H rules were added to the in situ histology section.

2. Cancer PathCHART Specialty Matter Expert review of breast histologies determined some histologies with individual ICD-O codes are to be considered synonyms for the NOS term for cases diagnosed 1/1/2024 forward. Therefore, they have been moved from the subtype/variant column 3 to synonym column 2. These terms have been identified with the symbol **++.** Terms and codes which were moved to column 2 are still listed in column 3 with the corresponding ICD-O code and note indicating valid for cases diagnosed prior to 1/1/2024.

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with; (duct **and** lobular is equivalent to duct **with** lobular)
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Behavior code /2; DCIS; intraductal; noninfiltrating; noninvasive; carcinoma in situ
- Carcinoma; adenocarcinoma
- De novo; new tumor; frank (obsolete term)
- Duct; ductal; NST (no special type); carcinoma NST; mammary carcinoma
- Mammary; breast
- Simultaneous; synchronous; existing at the same time; concurrent; prior to first course treatment
- Topography; site code
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician's statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to **determine** multiple primaries
  - **Do not** use these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

Terms that are NOT Equivalent or Equal

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

- **Phenotype** is not equivalent to **subtype/type/variant**
- **Invasive carcinoma, NST with lobular features** is not equivalent to **invasive carcinoma with ductal and lobular features**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 1: Primary Site Codes

Table 1 contains terms used in **mammograms, clinical diagnosis,** and less frequently the **operative and pathology reports** to describe the **location** of the tumor. Find the **term** in Column 1 and use the **site code** in Column 2.

Note: See the **"clock" diagram** at the end of the Equivalent Terms and Definitions for a graphic of the o'clock designations and corresponding **quadrants/subsites** of the breast.

Refer to the **SEER Manual** and **COC Manual** for a **priority list** for using documents such as mammograms, operative reports, and pathology reports to determine the tumor location.

Column 1 includes terms used to describe the **location/site** of the tumor.
Column 2 contains the **site term and code.**

Terms and Descriptive Language	Site Term and Code
Areolar Nipple Paget disease **without** underlying tumor ***Note:*** Paget with underlying tumor is coded to the quadrant of breast in which the underlying tumor is located	Nipple **C500**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Terms and Descriptive Language	Site Term and Code
Above nipple Area extending 1 cm around areolar complex Behind the nipple Below the nipple Beneath the nipple Central portion of breast Cephalad to nipple Infra-areolar Lower central Next to areola NOS Next to nipple Retroareolar Subareolar Under the nipple Underneath the nipple	Central portion of breast **C501**
Superior inner Superior medial Upper inner quadrant (UIQ) Upper medial	Upper inner quadrant of breast **C502**
Inferior inner Inferior medial Lower inner quadrant (LIQ) Lower medial	Lower inner quadrant of breast **C503**
Superior lateral Superior outer Upper lateral Upper outer quadrant (UOQ)	Upper outer quadrant of breast **C504**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Terms and Descriptive Language	Site Term and Code
Inferior lateral Inferior outer Lower lateral Lower outer quadrant (LOQ)	Lower outer quadrant of breast **C505**
Axillary tail of breast Tail of breast NOS Tail of Spence	Axillary tail of breast **C506**
12:00 o'clock 3:00 o'clock 6:00 o'clock 9:00 o'clock Inferior breast NOS Inner breast NOS Lateral breast NOS Lower breast NOS Medial breast NOS Midline breast NOS Outer breast NOS Overlapping lesion of breast Superior breast NOS Upper breast NOS	Overlapping lesion of breast C**508** ***Note:*** This is a **single tumor** which **overlaps quadrants/subsite.**
¾ or more of breast involved with tumor Diffuse (tumor size 998) Entire breast Inflammatory without palpable mass Multiple tumors in different subsites (quadrants) within the same breast	Breast NOS **C509** ***Note:*** Used for: • Non-contiguous **multiple** tumors in **different quadrants/subsites** of same breast **OR** • **Unknown/unable to identify** in which **quadrant/subsite** the tumor is l**ocated** (Example: Outpatient biopsy with no quadrant identified. Patient lost to follow-up.) • Inflammatory carcinoma; diffuse tumor

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 2: Histology Combination Codes

Instructions:

1. Use Table 2 when instructed to by the Multiple Primary and Histology Rules.
2. Compare the **terms** in the **diagnosis** (pathology, cytology, radiographic, clinical) to the terms in **Column 1.**
3. When the terms **match**, use the **combination code** listed in **Column 2.**
4. The **last row** in the table is a **"last resort"** code: adenocarcinoma mixed subtypes **8255**.
5. Use the combination codes only when the histologies are in a **single** tumor **OR multiple tumors** abstracted as a single primary.
6. Mixed histologies may be described as follows:
   A. A "**combination** of"
   B. Histology 1 **AND** histology 2
   C. Histology 1 **WITH** histology 2
   D. **Mixed** histology 1 and histology 2

Note 1: **Do not** use Table 2 in the following situations:

- For tumors with both **invasive** and **in situ** behavior. The **Histology Rules** instruct to code the invasive histology.
- When one of the histologies is described as **differentiation or features**
- When the terms are a **NOS** and a **subtype/variant** of that NOS. See the **Histology Rules** for instructions on coding a NOS and a subtype/variant in a single tumor or multiple tumors abstracted as a single primary.

Note 2: Some histologies can be in situ or invasive; others are limited to either /2 or /3 behavior code.

- When a code is **limited to in situ, /2** will be **added** to the code (both components are in situ)
- When a code is **limited to invasive, /3** will be **added** to the code (both components are invasive)

Note 3: This table is not a complete listing of histology combinations.

Column 1 contains the required ICD-O histology terms.
Column 2 contains the histology **combination term** and **code.**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Required Histology Terms	Histology Combination Term and Code
DCIS/duct carcinoma/carcinoma NST **8500** **AND** LCIS/lobular carcinoma **8520 or 8519** ***Note 1:*** Histologies may be a mix of in situ and invasive ***Note 2:*** 8522 is used when: • Duct and lobular carcinoma are present in a single tumor **OR** • Duct is present in at least one tumor and lobular present in a least one tumor in the same breast **OR** • One tumor is mixed duct and lobular; the other tumor in the same breast is either duct or lobular **OR** • All tumors in the same breast are mixed duct and lobular ***Example:*** One tumor with invasive duct carcinoma in LOQ RT breast; second tumor with invasive lobular carcinoma in UOQ RT breast ***Note 3:*** **Do not** use 8522 when the diagnosis is carcinoma NST/duct carcinoma with lobular **differentiation.** See Histology Rules for instructions on **coding differentiation**.	DCIS and in situ lobular carcinoma **8522/2** Note: The lobular includes pleomorphic lobular carcinoma in situ **8519/2** Invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma **8522/3** ***Note 1:*** CAP uses the term Invasive carcinoma with ductal and lobular features ("mixed type carcinoma") to indicate both duct and lobular are present. ***Note 2:*** This is an exception to the instruction that features are not coded. ***Note 3:*** Carcinoma NST includes all subtypes of carcinoma NST ***Note 4:*** Lobular carcinoma includes invasive pleomorphic lobular carcinoma **Additional combinations of duct and lobular coded 8522/3**: • Intraductal and lobular carcinoma (includes invasive pleomorphic lobular carcinoma) • Infiltrating duct and lobular carcinoma in situ (LCIS) • Infiltrating duct and pleomorphic lobular carcinoma in situ • Infiltrating lobular carcinoma and ductal carcinoma in situ (DCIS) Infiltrating pleomorphic lobular carcinoma and ductal carcinoma in situ (DCIS)

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Required Histology Terms	Histology Combination Term and Code
DCIS/duct carcinoma/carcinoma NST **OR** any ONE subtype/variant of carcinoma NST **AND** **Any** histology in **Table 3 with exception** of • Lobular carcinoma **8520** and pleomorphic lobular carcinoma in situ **8519/2*** • Paget disease **8540** ***Note 1:*** Both histologies **must have** the **same behavior** code. ***Note 2:*** See **Table 3** for carcinoma NST/duct carcinoma subtypes/variants. ***Note 3:*** **Do not** use combination code for duct with lobular **differentiation.** This is a synonym for carcinoma NST.	Invasive carcinoma NST/duct mixed with other types of invasive carcinoma **8523/3** DCIS mixed with other in situ carcinoma **8500/2** ***Note***: Prior to 2018, DCIS and other in situ was coded **8523/2**.
Lobular carcinoma **AND** **Any** histology in **Table 3 with exception** of • Duct carcinoma/carcinoma NST/DCIS (and subtypes/variants) **8500** • Paget disease, in situ and invasive ***Note 1:*** See **Table 3** for carcinoma NST/duct carcinoma subtypes/variants. ***Note 2:*** This code does not include lobular and Paget disease. See Multiple Primary Rules. Lobular carcinoma and Paget are separate primaries.	Infiltrating lobular mixed with other types of carcinoma **8524/3** In situ lobular mixed with other types of in situ carcinoma **8524/2** **(Cases diagnosed prior to 1/1/2024 only)** ***Note***: Beginning with cases diagnosed 1/1/2024 forward, in situ lobular carcinoma with other types of in situ carcinoma 8524/2 has been deemed biologically impossible based on expert pathologist review for the Cancer PathCHART project. See Rule H7 and H8 for coding instructions.

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Required Histology Terms	Histology Combination Term and Code
Metaplastic carcinoma OR any ONE subtype/variant of metaplastic carcinoma **AND** **Duct carcinoma/carcinoma NST OR** **Lobular carcinoma**	Code metaplastic carcinoma **8575** OR Subtype/variant of metaplastic carcinoma **Note**: Metaplastic carcinoma, NOS and subtypes are almost always mixed with invasive mammary carcinoma, NST and at times lobular carcinoma. These tumors should be coded to metaplastic regardless of percent invasive mammary carcinoma or lobular carcinoma present.
Paget disease **AND** **Underlying** DCIS ***Note:*** Paget disease is classified as malignant /3 in the ICD-O. Paget disease is coded as in situ /2 **ONLY** when the pathology states the Paget disease is in situ.	Paget disease (invasive or behavior not specified) and DCIS/intraductal carcinoma **8543/3** **Paget disease** (specified as in situ) and DCIS/intraductal carcinoma **8543/2**
Paget disease **AND** **Underlying** infiltrating **duct** carcinoma/carcinoma NST and **all subtypes/variants** of infiltrating duct/carcinoma NST (must be a /3) ***Note:*** See **Table 3** for subtypes/variants of carcinoma NST/duct carcinoma.	Paget disease and infiltrating duct carcinoma **8541/3**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Required Histology Terms	Histology Combination Term and Code
Any two invasive carcinoma NST subtypes/variants (percentage not stated) abstracted as a single primary ***Note 1:*** The diagnosis may be two subtypes/variants and the pathologist may mention the presence of duct/carcinoma NST. Ignore the mention of carcinoma NST. ***Note 2:*** See **Table 3** for subtypes/variants of carcinoma NST/duct carcinoma.	Adenocarcinoma with mixed subtypes **8255/3**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

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

Use Table 3 as directed by the **Histology Rules** to assign the more common histology codes for breast tumors.

Note 1: Rare histologies may not be listed in the table. When a histology term is not found, reference ICD-O and all updates.
Note 2: Submit a question to **Ask a SEER Registrar** when the histology is not found in Table 3, ICD-O or all updates.
Note 3: Behavior codes are listed when the term has only one possible behavior (either a /2 or /3). For histologies which may be either /2 or /3, a behavior code is not listed. Code behavior from pathology.
Note 4: Only use the histology code from the table when the diagnosis is **EXACTLY** the term listed.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including rhabdomyosarcoma 8900/3 (column 3). Rhabdomyosarcoma is also a NOS because it has subtypes/variants. The subtypes/variants are indented under the NOS (rhabdomyosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that rhabdomyosarcoma has subtypes/variants.
When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, rhabdomyosarcoma **8900/3** and alveolar type rhabdomyosarcoma **8920/3** are a NOS and a subtype/variant, **NOT** two different subtypes.

Table begins on next page

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Acinic cell carcinoma 8550	Acinar adenocarcinoma Acinar carcinoma	
Adenoid cystic carcinoma (ACC) 8200	ACC Adenocystic basal cell carcinoma Carcinoma adenoides cysticum Cylindromatous carcinoma	
Adenomyoepithelioma with carcinoma 8983	AME Malignant AME	
Apocrine carcinoma 8401 ***Note:*** This is a diagnosis that is **EXACTLY** apocrine **carcinoma**, **not** a carcinoma **NST** with apocrine **features**, **differentiation**, **or type.**		
Carcinoma NST 8500 ***Note***: Cribriform carcinoma may consist of up to 50% tubular formations. The term cribriform/tubular carcinoma is coded as cribriform carcinoma.	Carcinoma, NOS Carcinoma of no special type (ductal/NST) Carcinoma/carcinoma NST with choriocarcinomatous features Carcinoma/carcinoma NST with cribriform features Carcinoma/carcinoma NST with melanotic features Carcinoma/carcinoma NST with neuroendocrine features Carcinoma/carcinoma NST with signet ring cell differentiation DCIS **8500/2** DCIS of high nuclear grade **8500/2**	Carcinoma with osteoclastic-like stromal giant cells **8035** Cribriform carcinoma/Ductal carcinoma, cribriform type **8201/3;** Cribriform carcinoma in situ **8201/2** Pleomorphic carcinoma **8022/3** Ductal carcinoma in situ, solid type/intraductal carcinoma, solid type **8230/2** Solid carcinoma/solid adenocarcinoma **8230/3 ++(cases diagnosed prior to 1/1/2024 only)**

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
	DCIS of intermediate nuclear grade **8500/2** DCIS of low nuclear grade **8500/2** Duct/ductal carcinoma Duct/ductal carcinoma in situ **8500/2** Duct/ductal carcinoma NOS Duct/ductal carcinoma NST (no special type) Duct/ductal carcinoma with apocrine features Duct/ductal carcinoma with apocrine metaplasia Duct/ductal carcinoma with lobular features Duct/ductal carcinoma with micropapillary features Duct/ductal carcinoma with mucin production Duct/ductal carcinoma with neuroendocrine features Duct/ductal carcinoma with squamous metaplasia Infiltrating ductal carcinoma **8500/3** Intraductal carcinoma **8500/2** Invasive carcinoma with medullary features **8500/3** Invasive carcinoma with micropapillary features **8500/3** Invasive carcinoma with neuroendocrine features **8500/3** Invasive carcinoma not otherwise specified (ductal/NOS) **8500/3** Invasive carcinoma NST with metaplastic features **8500/3**	

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
	Invasive carcinoma NST/duct with medullary features **8500/3** Invasive carcinoma, with signet-ring cell features **8500/3** Invasive carcinoma of no special type (NST) **8500/3** Invasive carcinoma with clear cell (glycogen rich) features **8500/3** Invasive carcinoma, NST **8500/3** Invasive carcinoma, type cannot be determined **8500/3** Invasive mammary carcinoma **8500/3** Invasive mammary carcinoma associated with encysted papillary carcinoma **8500/3** Invasive mammary carcinoma NST with lobular features **8500/3** Invasive mammary carcinoma NST with medullary features **8500/3** Invasive mammary carcinoma NST with mucinous features **8500/3** Invasive mammary carcinoma NST with neuroendocrine features **8500/3** Invasive mammary carcinoma NST with tubulo-lobular variant **8500/3** Invasive mammary carcinoma with apocrine features **8500/3** Invasive mammary carcinoma with cribriform features **8500/3**	

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
	Invasive mammary carcinoma with tubular features **8500/3** Invasive solid carcinoma/ adenocarcinoma **8500/3 ++(cases diagnosed 1/1/2024 forward)** Mammary carcinoma in situ **8500/2** Mammary carcinoma/cancer Non-invasive mammary carcinoma **8500/2**	
Glycogen-rich clear cell carcinoma 8315	Glycogen-rich carcinoma	Clear cell carcinoma **8310**
Inflammatory carcinoma 8530		
Lipid-rich carcinoma 8314	Lipid-secreting carcinoma	
Lobular carcinoma 8520	Alveolar lobular carcinoma Classic lobular carcinoma Florid lobular carcinoma **8520/2** Intraductal papilloma with lobular carcinoma in situ **8520/2** Invasive lobular carcinoma, alveolar type/variant **8520/3** Invasive lobular carcinoma, solid type **8520/3** Lobular carcinoma in situ **8520/2** Lobular carcinoma with cribriform features Mixed lobular carcinoma (lobular carcinoma NOS and one or more variants of lobular carcinoma) Invasive pleomorphic lobular carcinoma **8520/3** Solid lobular carcinoma Tubulolobular carcinoma	Pleomorphic lobular carcinoma in situ **8519/2*** ***Note:*** 8519/2 is a new code for in situ /2 tumors only.

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Medullary carcinoma 8510	MC	Atypical medullary carcinoma (AMC) **8513**
Metaplastic carcinoma NOS or of no special type (NST) 8575 ***Note 1*:** Squamous cell carcinoma of the breast is **extremely rare.** Carefully **check** the **pathology** report to verify the squamous cell originated in the breast parenchyma, rather than the skin of the breast. ***Note 2*:** Metaplastic carcinoma, NOS and subtypes are almost always mixed with invasive mammary carcinoma, NST and at times lobular carcinoma. These tumors should be coded to metaplastic regardless of percent invasive mammary carcinoma or lobular carcinoma present.	Invasive mammary carcinoma with matrix production Metaplastic carcinoma, mixed epithelial and mesenchymal type Metaplastic carcinoma with mesenchymal differentiation Metaplastic carcinoma with squamous features Metaplastic carcinoma with other types of mesenchymal differentiation Mixed metaplastic carcinoma Metaplastic carcinoma spindle- cell type/spindle cell carcinoma ++**(cases diagnosed 1/1/2024 forward)**	Carcinosarcoma **8980/3** Fibromatosis-like metaplastic carcinoma **8572** Low grade adenosquamous carcinoma **8560** Metaplastic carcinoma spindle-cell type/spindle cell carcinoma **8032 ++(cases diagnosed prior to 1/1/2024)** Metaplastic carcinoma with chondroid differentiation/with osseous differentiation **8571** Myoepithelial carcinoma **8982** Squamous cell carcinoma **8070**

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Mucinous carcinoma 8480 *Note 1:* This is a diagnosis that is **EXACTLY** "mucinous carcinoma," "mucinous duct carcinoma," "mucinous DCIS" OR "greater than 90% mucinous." **See Histology Rules**. *Note 2:* Mucinous duct carcinoma is listed on the CAP protocol. It is not recognized by WHO or IARC. Mucinous carcinoma is not a subtype/variant of Carcinoma NST/duct carcinoma.	Colloid carcinoma Mucinous adenocarcinoma Mucoid carcinoma	
Mucoepidermoid carcinoma 8430		
Oncocytic carcinoma 8290		
Paget disease of the nipple with no underlying tumor 8540		
Papillary carcinoma 8503	Intraductal papillary carcinoma **8503/2*** Intraductal papillary carcinoma with DCIS **8503/2*** Intraductal papilloma with ductal carcinoma in situ **8503/2** Invasive ductal papillary carcinoma **8503/3** Invasive papillary carcinoma **8503/3** Papillary carcinoma of breast, NOS **8503/3** Papillary carcinoma non-invasive **8503/2*** Papillary ductal carcinoma in situ **8503/2***	Encapsulated papillary carcinoma, NOS/non-infiltrating/intracystic **8504/2** with invasion **8504/3** with invasive carcinoma, NST/invasive duct carcinoma **8504/3** Micropapillary carcinoma **8507*** Tall cell carcinoma with reverse polarity **8509/3;** Solid papillary carcinoma in situ **8509/2*** with invasion **8509/3***

Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Phyllodes tumor, malignant 9020/3	Cystosarcoma phyllodes, malignant Periductal stromal tumor, low grade	
Polymorphous carcinoma 8525		
Sarcoma NOS 8800/3 ***Note 1:*** Angiosarcoma **9120/3** is also a NOS with the following subtypes/variants: Lymphangiosarcoma 9170/3 Malignant hemangioendothelioma 9130/3 ***Note 2*:** Rhabdomyosarcoma **8900/3** is also a NOS with the following subtypes/variants: Alveolar type rhabdomyosarcoma **8920/3** Embryonal type rhabdomyosarcoma **8910/3** Pleomorphic rhabdomyosarcoma **8901/3** ***Note 3:*** Angiosarcoma has the following synonyms (they are not subtypes/variants): Epithelioid angiosarcoma Hemangiosarcoma Post radiation angiosarcoma of breast		Angiosarcoma **9120/3** Epithelioid angiosarcoma Hemangiosarcoma Post radiation angiosarcoma of breast Lymphangiosarcoma **9170/3** Malignant hemangioendothelioma **9130/3** Liposarcoma **8850/3** Leiomyosarcoma **8890/3** Osteosarcoma **9180/3** Rhabdomyosarcoma **8900/3** Alveolar type **8920/3** Embryonal type **8910/3** Pleomorphic **8901/3**

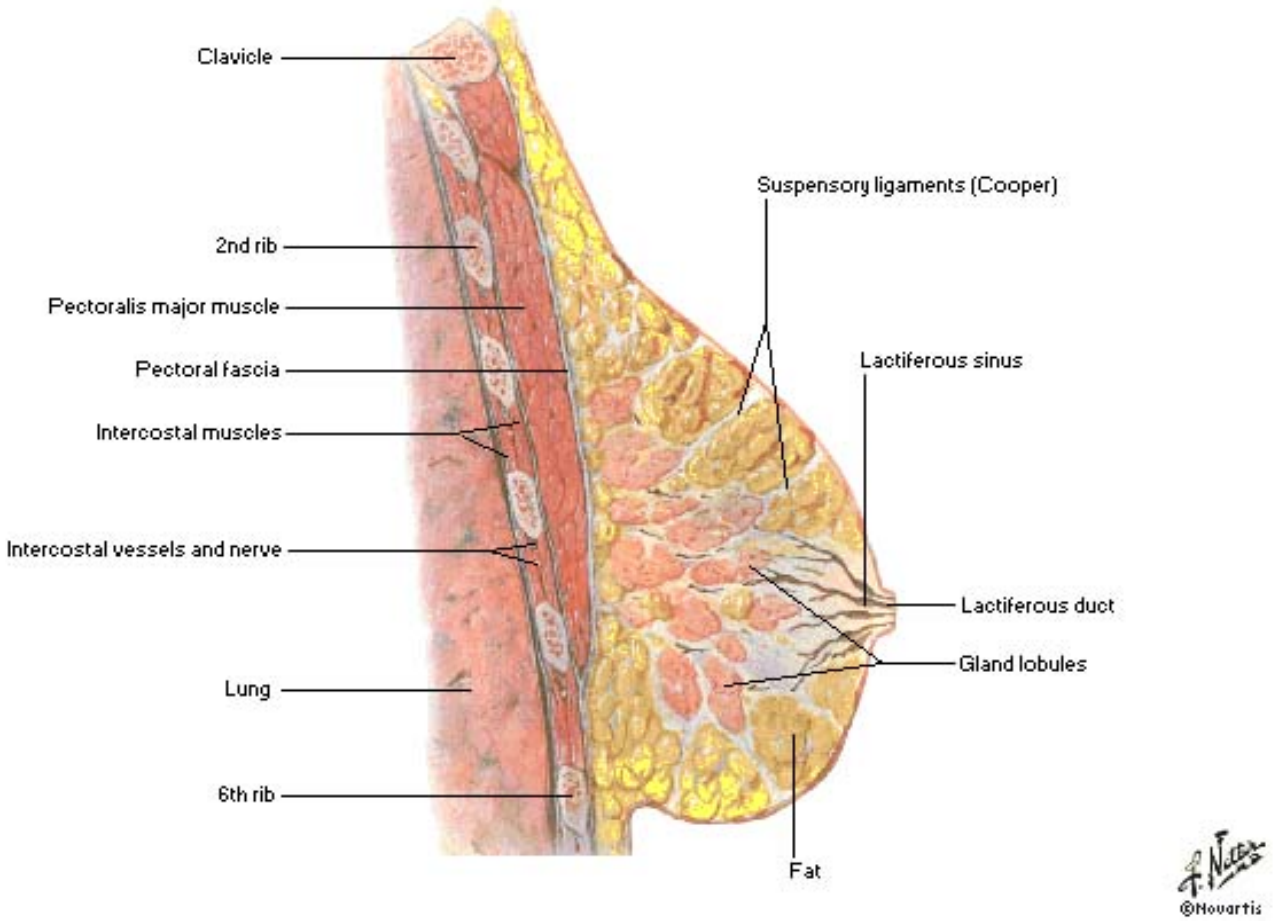
Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Sebaceous carcinoma 8410		
Secretory carcinoma 8502	Juvenile breast carcinoma	
Signet ring carcinoma 8490		
Small cell carcinoma 8041	Carcinoid tumor of breast Endocrine carcinoma Neuroendocrine carcinoma, poorly differentiated	Carcinoma with neuroendocrine differentiation **8574/3** Neuroendocrine tumor, well-differentiated **8246/3**
Tubular carcinoma 8211		

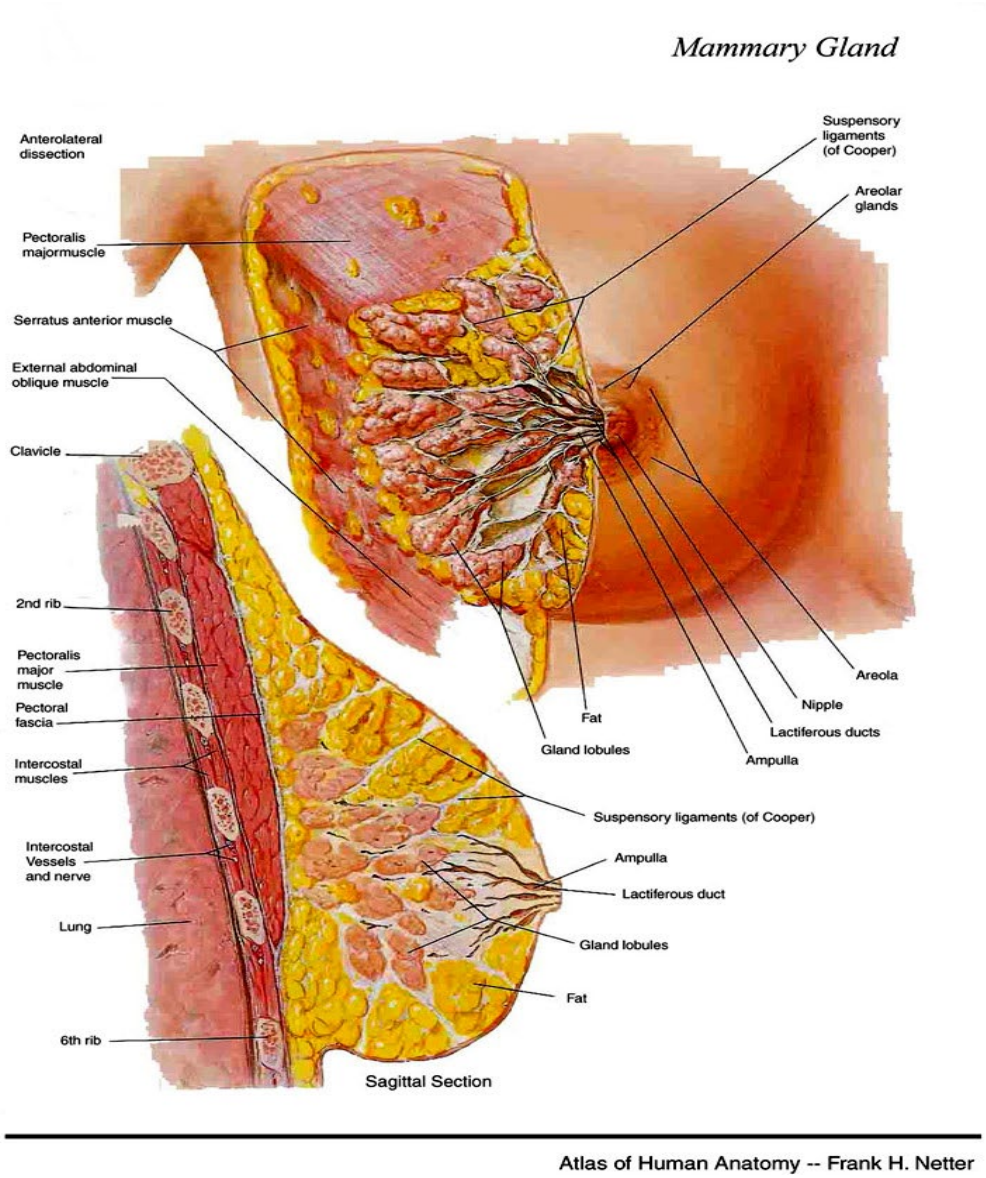
***New codes approved by IARC/WHO Committee for ICD-O**

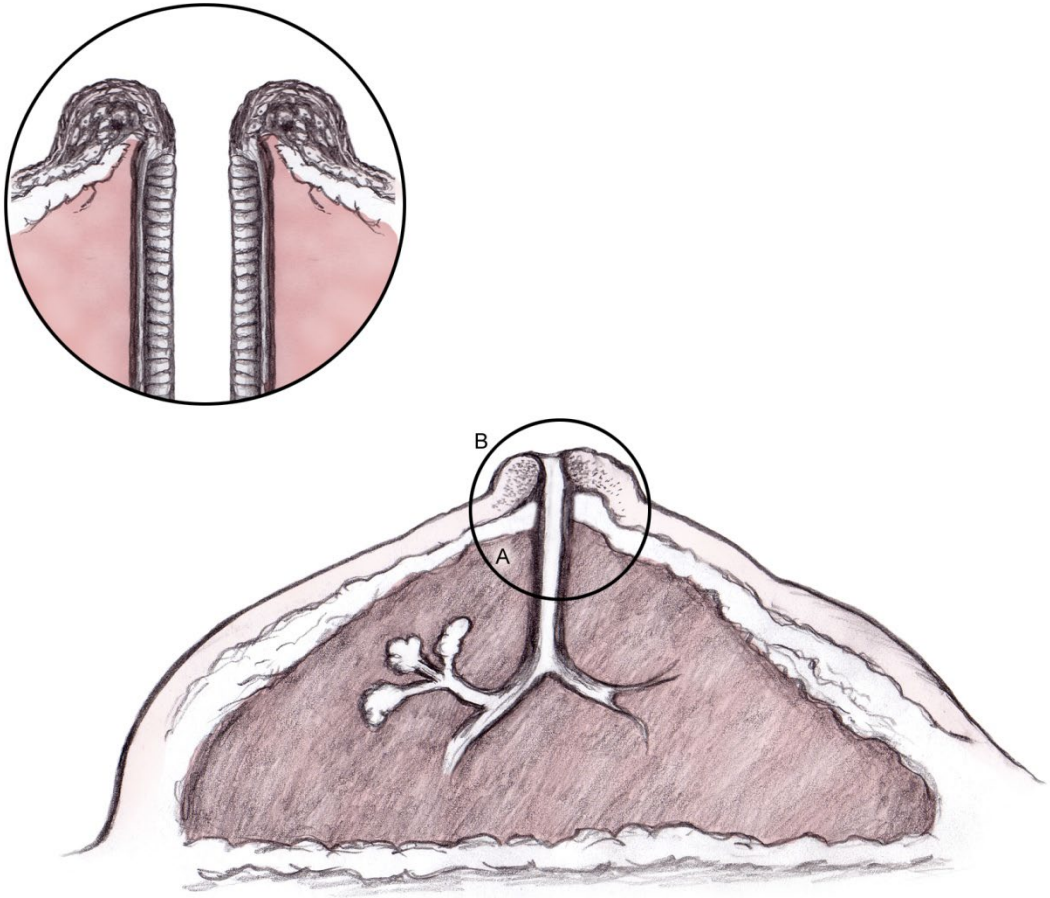
++Denotes change per Cancer PathCHART Specialty Matter Expert review. ICD-O codes and terms with ++ have a separate ICD-O code, however, per the expert review, are considered a synonym for the NOS term. This change applies to cases diagnosed 1/1/2024 forward. The terms and ICD-O codes remain subtype/variants for cases diagnosed prior to 1/1/2024.

Illustrations



Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)





Paget Disease of the nipple. Shows growth pattern of Paget on the pigmented portion of nipple and inside the milk duct opening

Source:
*"Image reprinted with permission from eMedicine.com, 2010. Available at: **http://emedicine.medscape.com/article/1101235-overview***

Breast Equivalent Terms and Definitions
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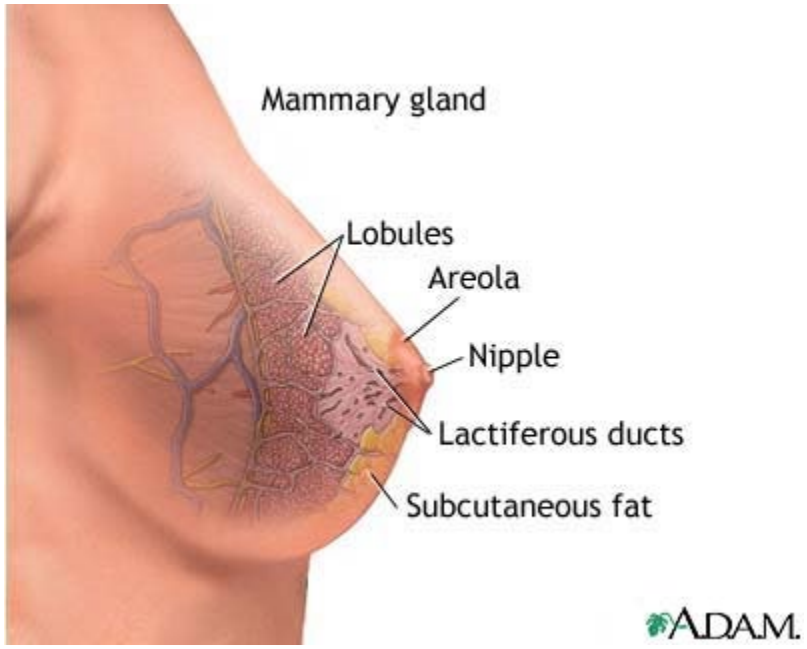
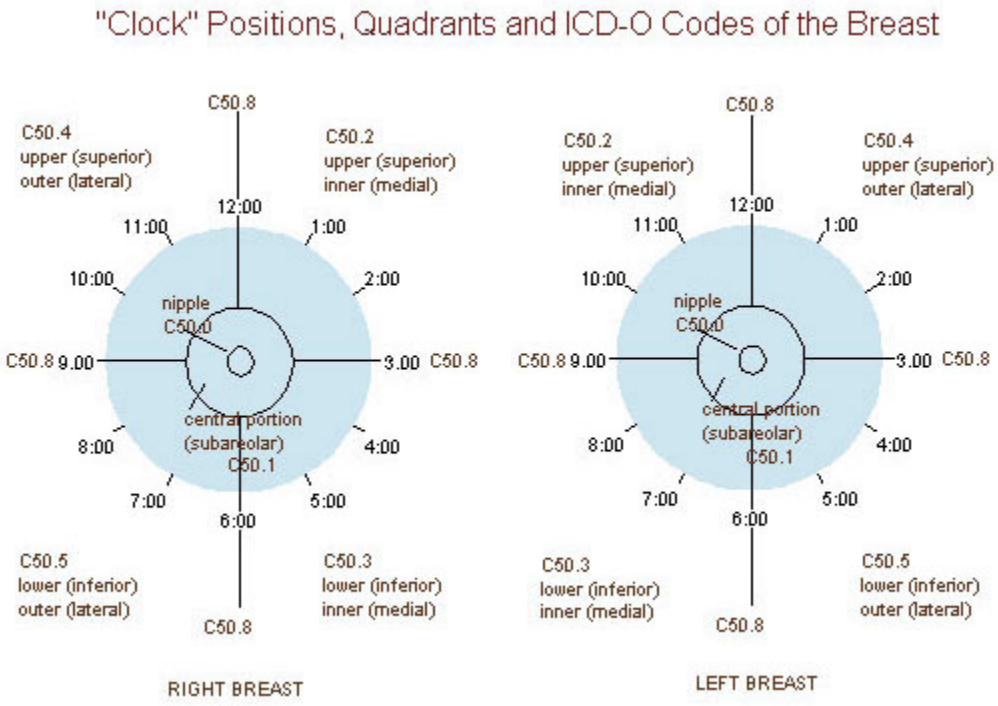
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

The position of the tumor in the breast may be described as the positions on a clock

The two circles in the graphic are
Innermost circle: Retroareolar (under/behind areola)
Outer circle: Central portion of breast



Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Axillary lymph nodes
- Bone
- Brain
- Chest wall
- Discontinuous involvement of skin of breast
- Distant lymph nodes as identified in Summary Staging Manual
- Liver
- Lung

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include:

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors

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

Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

IMPORTANT: If the current tumor was **preceded** by a tumor in the same breast or contralateral breast, go to the **Multiple Tumors module.**

Rule M2 Abstract a **single primary**[i] when the diagnosis is **inflammatory carcinoma** in:

- Multiple quadrants of same breast **OR**
- Bilateral breasts

Rule M3 Abstract a **single primary**[i] when there is a **single tumor**.

Note 1: A single tumor is always a single primary.
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites/quadrants.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor

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

Multiple Tumors

Note 1: Multiple tumors may be single primary or multiple primaries**.**
***Note 2*:** ER, PR, and/or HER2 are not used to determine multiple primaries.
Note 3: A subsequent tumor in the chest wall or surgical scar **without** evidence of residual breast tissue is regional metastasis.

Rule M4 Abstract **multiple primaries**[ii] when there are **separate**, **non-contiguous** tumors in sites with ICD-O site codes that **differ** at the second (C**X**xx) and/or third characters (Cx**X**x).

Note 1: Tumors with site codes that differ at the second or third character are in **different primary sites;** for example, a breast tumor C**50x** and a colon tumor C**18**x differ at the second and third character.
Note 2: This rule **does not** include metastases. Metastatic tumors are not used to determine multiple primaries; for example, liver metastases from the breast cancer would not be counted as a second primary.

Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M5 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being **clinically disease-free** for **greater than five years** after the original diagnosis or last recurrence.

Note 1: The rules are hierarchical. This rule **only** applies when there is a **subsequent tumor in the same breast.** In other words, a primary in the contralateral breast does not start the "clock" over.

Note 2: **Clinically** disease-free means that there was **no evidence** of recurrence on follow-up.

- Mammograms are NED
- Scans are NED

Note 3: When there is a recurrence less than or equal to five years of diagnosis, the **"clock"** starts over. The time interval is calculated from the **date of last recurrence.** In other words, the patient must have been disease-free for greater than five years from the date of the last recurrence.

Note 4: When it is **unknown/not documented** whether the patient had a recurrence, use **date of diagnosis** to compute the time interval.

Note 5: The physician may state this is a **recurrence,** meaning the patient had a previous breast tumor and now has another breast tumor. **Follow the rules;** do not attempt to interpret the physician's statement.

Note 6: When a breast resection was done and a subsequent tumor is identified in the remaining chest wall, muscle, or skin AND there was no residual breast tissue identified in the resected specimen, this is a recurrence and not a new primary.

Rule M6 Abstract a **single primary**[i] when there is **inflammatory carcinoma** in:

- Multiple quadrants of same breast **OR**
- Bilateral breasts

Rule M7 Abstract **multiple primaries**[ii] when there is **bilateral** breast cancer (both right and left breast).

Note 1: Physician statement of "bilateral breast cancer" should **not be interpreted** as meaning a single primary. The term is not used consistently. The literal definition of bilateral is "cancer in both breasts".

Note 2: The histologies within each breast may be the same or different.

Rule M8 Abstract a **single primary**[i] when the diagnosis is **Paget disease with synchronous underlying** in situ or invasive carcinoma NST (duct/ductal) or subtypes of duct.

Note: If the underlying tumor is any histology **other than** duct or subtypes of duct, continue through the rules.

Rule M9 Abstract **multiple primaries**[ii] when the diagnosis is **Paget disease with underlying** tumor which is NOT duct.

Example: Paget disease of the nipple with underlying lobular carcinoma are multiple primaries.

Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M10 Abstract a **single primary**[i] when there are multiple tumors of **carcinoma NST/duct and lobular**.

- Both/all tumors may be a mixture of carcinoma NST/duct and lobular 8522 **OR**
- One tumor may be duct and another tumor lobular **OR**
- One tumor may be mixed duct and lobular 8522, the other tumor either duct or lobular

Note 1: Tumors must be in the same breast.

Note 2: Carcinoma NST/duct includes:

- DCIS **8500/2**
- Carcinoma NST **8500/3**
- Carcinoma with osteoclastic-like stromal giant cells **8035/3** (subtype/variant of carcinoma NST)
- Cribriform carcinoma **8201/3**
- Pleomorphic carcinoma **8022/3**

Note 3: Lobular carcinoma includes:

- In situ lobular carcinoma **8520/2**
- In situ pleomorphic lobular carcinoma **8519/2**
- Invasive lobular carcinoma **8520/3**
- Invasive pleomorphic lobular carcinoma **8520/3**

Note 4: **When a mixture of behaviors is present in carcinoma, NST and lobular carcinoma, follow the H rules to determine the correct histology code.**

Note 5: For cases initially diagnosed as in situ with subsequent invasive tumor and stated to be a single primary per M10, edit the original abstract as follows:

- Do not change date of diagnosis.
- For cases which have been abstracted, change behavior code on original abstract from /2 to 8522/3.
- Report all data changes for cases which have been submitted to the central registry.

Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M11 Abstract a **single primary**[i] when a **ductal** carcinoma occurs **after a combination code** in the same breast. See the following list:

- **DCIS** following a diagnosis of:
  - DCIS + lobular carcinoma in situ **8522/2 OR**
  - DCIS + in situ Paget **8543/2 OR**
  - DCIS + Invasive Paget **8543/3 OR**
  - DCIS mixed with other in situ **8523/2** (code used for cases diagnosed prior to 1/1/2018)
- **Invasive carcinoma NST/duct** following a diagnosis of:
  - Invasive duct + invasive lobular **8522/3 OR**
  - Invasive duct + invasive Paget **8541/3 OR**
  - Invasive duct + other invasive carcinoma **8523/3**

Rule M12 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3 of **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS**: Encapsulated papillary carcinoma with invasion 8504/3 and solid papillary carcinoma with invasion 8509/3 are both subtypes of invasive papillary carcinoma 8503/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Encapsulated papillary carcinoma 8504/2 is a subtype/variant of in situ papillary carcinoma 8503/2. Pleomorphic lobular carcinoma in situ 8519/2 is a subtype/variant of lobular carcinoma in situ 8520/2. They are distinctly different histologies. Abstract multiple primaries.

Rule M13 Abstract a **single primary**[i] when **synchronous**, separate/non-contiguous tumors are on the **same row** in **Table 3** in the Equivalent Terms and Definitions.

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3) **OR**
- A NOS histology in column 3 with an indented subtype/variant

Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M14 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are:

- On **different rows** in Table 3 in the Equivalent Terms and Definitions
- A combination code in Table 2 and a code from Table 3

Note 1: Timing is irrelevant. Tumors may be synchronous or non-synchronous.

Note 2: Each row in the table is a distinctly different histology.

Example 1: Paget disease of the nipple with underlying lobular are multiple primaries. Paget and lobular are on different rows in Table 3.

Example 2: Two tumors right breast. One tumor is invasive mixed duct and lobular 8522/3 (combination code from Table 2) and the second tumor is tubular 8211/3 (histology from Table 3). Abstract two primaries: 8522/3 and 8211/3.

Rule M15 Abstract a **single primary**[i] (the invasive) when an **in situ** tumor is diagnosed **after** an **invasive** tumor in the same breast.

Note 1: Once the patient has an invasive tumor, the **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.

Note 2: The rules are **hierarchical.** Only use this rule when none of the previous rules apply.

Note 3: The tumors may be a **NOS** and a **subtype/variant** of that NOS.

Rule M16 Abstract a **single primary**[i] (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60** days **after** an **in situ** tumor in the same breast.

Note 1: The rules are **hierarchical.** Only use this rule when none of the previous rules apply.

Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS.

Note 3: When the case has been abstracted, **change behavior** code on original abstract from /2 to /3.

Note 4: Do **not** change **date of diagnosis**.

Note 5: If the case has already been submitted to the central registry, **report** all changes.

Note 6: The physician **may stage both** tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

Note 7: See the **COC** and **SEER manuals** for instructions on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Breast Multiple Primary Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M17 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor in the same breast.

Note 1: The rules are **hierarchical.** Only use this rule when none of the previous rules apply.

Note 2: Abstract **both** the invasive and in situ tumors.

Note 3: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression.

Note 4: This rule is based on long-term **epidemiologic** studies of recurrence intervals. The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were also authors, co-authors, or editors of the **AJCC** Staging Manual.

Rule M18 Abstract a **single primary**[i] when none of the previous rules apply.

Note: Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

Example: One tumor is invasive carcinoma NST/ductal 8500/3 and a separate non-contiguous tumor in the same breast is DCIS 8500/2. Abstract a single primary: invasive carcinoma NST/ductal 8500/3.

This is the end of instructions for Multiple Tumors.

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[i] Prepare one abstract. Use the histology rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

Note: Only code **differentiation** or **features** when there is a **specific code** for the NOS with differentiation or the NOS with features in **Table 2** **or** **Table 3** or the ICD-O and all updates.

Breast Histology Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Coding Histology

Note 1: The rules for coding breast histology are different from the histology coding rules for all other sites. **DO NOT USE THESE RULES FOR ANY SITE OTHER THAN BREAST.**
Note 2: Only use this section for one or more histologies within a single tumor.
Note 3: Do not use this section in place of the Histology Rules.

Two INVASIVE histologies

Two histologies within a single tumor will be either:

- A NOS and a subtype/variant **OR**
- Different histologies (different rows in Table 3 **OR** different subtypes in Table 3 Column 3 **OR** a combination code from Table 2 and a code from Table 3)

The following instructions are in priority order:

1. **NOS and a subtype/variant**
   A. Code the **subtype/variant** (specific histology) **ONLY** when documented to be **greater than 90%** of the tumor.
      Note: When a histology is listed as "minimal", "focus/foci/focal", "microscopic", you can assume the other histological portion comprises greater than 90% of the tumor.
      Example: Patient had an excisional biopsy with a pathologic diagnosis of invasive cribriform carcinoma 8201/3. There was microscopic involvement of one margin. The patient chose to have a total mastectomy. Pathology from the total mastectomy showed minimal residual invasive carcinoma NST 8500/3. Because the invasive carcinoma NST was minimal, the subtype/variant invasive cribriform carcinoma 8201/3 is assumed to be greater than 90% of the tumor.
   B. Code the **NOS/NST** when the subtype/variant is documented to be **less than or equal to 90%** of the tumor **OR** the percentage of subtype/variant is **unknown/not documented**.

2. Different histologies
   A. Code the histology which comprises the majority of tumor.
      Note 1: This instruction **does not apply** to:
      - Invasive carcinoma NST/ductal and lobular carcinoma (use the combination code 8522/3).
      - Mucinous carcinoma and a different histology (see Histology Rules)
      - Metaplastic carcinoma, NOS and subtypes/variants and invasive carcinoma, NST (see Histology Rules)

      Note 2: The following terms **do not** describe the majority of tumor.

Architecture	Pattern(s)
Component	Subtype
Differentiation*	Type
Features (of)*	Variant
Foci; focus, focal	

*Unless there is an exact ICD-O term that includes "differentiation" or "features"

   B. Code a combination code using **Table 2** in the Equivalent Terms and Definitions when the majority is unknown/not documented.

      Do not code **apocrine carcinoma** when the diagnosis specifies apocrine differentiation or features. **Apocrine differentiation** is frequently present in:
      - Carcinoma NST/duct carcinoma
        - Subtypes/variants of carcinoma NST/duct carcinoma
      - Lobular carcinoma NOS
        - Pleomorphic lobular carcinoma in situ

Ambiguous Terminology

3. Code the specific histology described by **ambiguous terminology** (list follows) **ONLY** when A or B is true:
   A. The only diagnosis available is **one histology** term described by ambiguous terminology
      - CoC and SEER require reporting of cases diagnosed only by ambiguous terminology

- Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

Example: Outpatient biopsy says probably apocrine carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology apocrine carcinoma. The case meets the criteria in #3A.

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

Example 1: The pathology diagnosis is carcinoma NST consistent with pleomorphic carcinoma. The oncology consult says the patient has pleomorphic carcinoma of the right breast. This is clinical confirmation of the diagnosis. Code pleomorphic carcinoma. The case meets the criteria in bullet 1.

Example 2: The pathology diagnosis is sarcoma consistent with liposarcoma. The treatment plan says the patient will receive the following treatment for liposarcoma of the breast. Treatment plan confirms liposarcoma. Code liposarcoma. The case meets the criteria in bullet 2.

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

List of Ambiguous Terminology

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

Breast Histology Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed **prior to neoadjuvant treatment.**
   Note 1: Histology changes do occur following immunotherapy, chemotherapy, hormone, 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, core biopsy, 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. For breast primaries, you cannot determine if histology comprises greater than 90% of the tumor by these diagnostic methods.

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

Use documentation in the following priority order to identify the histology type(s):

1. **Tissue or pathology report from primary site** (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol

   Note 1: Addendums and comments on the pathology report are given a high priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
   Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority. The final diagnosis is often the synoptic CAP report.
   Note 3: The CAP protocol is a checklist which:
   - Provides guidelines for collecting the essential data elements for complete reporting of malignant tumors and optimal patient care.
   - Allows physicians to check multiple histologies

2. **Cytology** (nipple discharge or fine needle aspirate (FNA) of primary site)

3. **Tissue/pathology from a metastatic site**
   Note 1: Code the behavior /3.
   Note 2: The tissue from a metastatic site often shows **variations** from the primary tumor. When it is the only tissue available, it is more accurate than imaging.

4. **Radiography:** The following list is **not in priority** order because they are not a reliable method for **identifying** specific **histology**(ies). They are, however, valuable in diagnosing a malignancy.
   A. Mammography
   B. Ultrasound
   C. CT
   D. MRI

5. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following **priority order:**
   A. Treatment Plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that refers to original pathology, cytology, or scan(s)
   D. Physician's **reference** to type of cancer (histology) in the medical record
   Note 1: Code the specific histology when documented.
   Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented.

Breast Histology Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor: In Situ Only

Note 1: DCIS is often multifocal/multicentric; use this module.
Note 2: Subtypes/variant, architecture, pattern, and features **ARE NOT CODED**. The majority of in situ tumors will be coded to DCIS 8500/2.

Rule H1 Code Paget disease in situ **8540/2** when the diagnosis is **exactly** Paget disease in situ.
Note 1: This is a **de novo** primary of the **nipple** (new tumor) with **no underlying** tumor.
Note 2: Paget is coded as in situ /2 **only** when **pathology documents** in situ behavior.

Rule H2 Code the histology when only **one histology** is present.
Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in **Table 3** and coding errors may occur if the table is not used.
Note 2: When the histology is **not listed** in **Table 3,** use the **ICD-O** and all **updates**.
Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 3, ICD-O or all updates.

Rule H3 Code DCIS and in situ lobular carcinoma **8522/2** when DCIS and in situ lobular carcinoma are present.
Note 1: Although the notes preceding the in situ section say most tumors will be coded to DCIS, 8522/2 identifies both DCIS and lobular carcinoma in situ.
Note 2: 8522/2 is the most accurate description of DCIS and lobular carcinoma in situ.
Note 3: 8522/2 includes DCIS and pleomorphic lobular carcinoma in situ.

Rule H4 Code DCIS and in situ Paget **8543/2.**
Note 1: Although the notes preceding the in situ section say most tumors will be coded to DCIS, 8543/2 identifies both DCIS and in situ Paget.
Note 2: 8543/2 is the most accurate description of DCIS and in situ Paget.

Rule H5 Code DCIS **8500/2** when there is a combination of DCIS and any other carcinoma in situ. See **Table 2**.

Rule H6 Code pleomorphic lobular carcinoma in situ **8519/2** when there is a combination of lobular carcinoma in situ and pleomorphic lobular carcinoma in situ.

Rule H7 Code in situ lobular carcinoma **8520/2** when there is a combination of lobular carcinoma in situ and one histology other than DCIS **AND**

- The percentage of lobular in situ comprises greater than 50% of the tumor **OR**
- Percentage of lobular in situ is unknown/not documented

Note: This is a new rule and applies to cases diagnosed 1/1/2024 forward. See H9 for cases diagnosed prior to 1/1/2024.

Rule H8 Code the histology that comprises greater than 50% of the tumor when two histologies are in situ lobular AND any histology other than DCIS.

Note: This is a new rule and applies to cases diagnosed 1/1/2024 forward. See H9 for cases diagnosed prior to 1/1/2024.

Rule H9 Code the histology using **Table 2** when there are multiple in situ histologies (2 or more) within a single tumor.

- Lobular and any histology other than DCIS **8524/2**
- Two or more histologies other than lobular and DCIS **8255/2**

Note: This rule does not include DCIS. See previous rules.

This is the end of instructions for a Single Tumor: In Situ Only

Code the histology according to the rule that fits the case

Single Tumor: Invasive and In Situ Components

Rule H10 Code the **invasive** histology when both invasive and in situ components are present (see Notes 2 and 3).

Note 1: **Ignore** the in situ term.

- This is consistent with the 2007 MPH Rules.

Note 2: When a single tumor has one of the histologies listed, see Table 3. These are specific histology terms that capture both invasive and in situ components.

- Encapsulated papillary carcinoma with invasion/with invasive carcinoma, NST/invasive duct carcinoma
- Solid papillary carcinoma with invasion

Note 3: When a single tumor has carcinoma NST/duct and lobular with different behaviors, continue through the rules.

Rule H11 Code duct and lobular **8522/3** when the final diagnosis is any of the following:

- Intraductal and invasive lobular carcinoma (includes invasive pleomorphic lobular carcinoma)
- Infiltrating duct and lobular carcinoma in situ (LCIS)
- Infiltrating duct and pleomorphic lobular carcinoma in situ
- Infiltrating lobular carcinoma and ductal carcinoma in situ (DCIS)
- Infiltrating pleomorphic lobular carcinoma and ductal carcinoma in situ (DCIS)

Note 1: Assign behavior code /3 even when an **in situ** histology is mixed with an **invasive**. This aligns with ICD-O-3.2 and was vetted with specialty matter experts.

Note 2: CAP uses the term **Invasive carcinoma with ductal and lobular features** ("mixed type carcinoma") as a synonym for duct carcinoma/carcinoma NST AND lobular carcinoma 8522/3.

Note 3: Although the instructions in the "Coding Multiple Histologies in a Single Tumor" section state, "Code the histology that comprises the majority of tumor", 8522/3 identifies both invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma and is the most accurate description.

This is the end of instructions for a Single Tumor: Invasive and In Situ Components

Code the histology according to the rule that fits the case

Breast Histology Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor: Invasive Only

Rule H12 Code Paget disease **8540/3** when the diagnosis is **exactly** Paget disease.

Note 1: This is a **de novo** primary of the **nipple** (new tumor) with **no underlying** tumor.

Note 2: Paget is coded /3 when:

- Pathology documents invasive behavior **OR**
- Behavior is not documented/unknown

Rule H13 Code the **underlying tumor** when there is a diagnosis of **inflammatory carcinoma**.[1]

Example: The patient has a clinical diagnosis of inflammatory breast carcinoma. Pathology shows carcinoma NST with dermal invasion as well as erythema. Code the underlying tumor: carcinoma NST 8500/3.

Informational item: The **clinical symptoms** of inflammatory breast cancer include rapid breast enlargement and skin changes (redness, edema peau d'orange) involving more than a third of the breast. Usually there is a diffuse firmness of the breast and there is no palpable underlying mass.

Note 1: Record the inflammatory carcinoma in **staging** fields.

Note 2: **Code inflammatory** carcinoma 8530/3 when it is the **only diagnosis** available (DCO, outpatient only, no follow-up).

Rule H14 Code mucinous carcinoma/adenocarcinoma **8480 ONLY** when:

- The diagnosis is **exactly** mucinous carcinoma or mucinous duct carcinoma **OR**
- Multiple histologies are present and mucinous carcinoma is documented as **greater than 90%** of the tumor

Note 1: The **pure** mucinous carcinoma category includes only cases which are diagnosed as exactly mucinous or documented to be greater than 90% of the tumor.

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

Note 3: When a tumor has both mucinous carcinoma and a different histology, and mucinous is less than or equal to 90% of the tumor (or the percentage is not documented), **code the other histology.**

[1] American College of Pathologists: **Protocol for the Examination of Specimens From Patients With Invasive Carcinoma of the Breast: "Inflammatory carcinoma requires the presence of clinical findings of erythema and edema involving at least one-third or more of the skin of the breast"**

Rule H15 Code the **primary invasive histology** when there is a carcinoma with **signet ring cells OR signet ring cell differentiation**.

Example: Resection pathology diagnosis is invasive lobular carcinoma with signet ring cell differentiation. Code the invasive lobular carcinoma **8520/3.**

Rule H16 Code **metaplastic carcinoma**, NOS, or subtype/variant of metaplastic carcinoma, NOS when **invasive carcinoma, NST OR invasive lobular carcinoma** is present along with the metaplastic carcinoma.

Example: Resection pathology diagnosis is invasive mammary carcinoma, NST with extensive metaplastic carcinoma present. Code metaplastic carcinoma **8575/3**.

Rule H17 Code the histology when only **one histology** is present.

Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in **Table 3** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Table 3,** use the **ICD-O** and all **updates**.

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

Rule H18 Code duct carcinoma and lobular carcinoma **8522/3** when the final diagnosis is invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma (includes invasive pleomorphic lobular carcinoma)

Note 1: CAP uses the term **Invasive carcinoma with ductal and lobular features** ("mixed type carcinoma") as a synonym for duct carcinoma/carcinoma NST AND lobular carcinoma 8522/3.

Note 2: Although the instructions in the "Coding Multiple Histologies in a Single Tumor" section state, "Code the histology that comprises the majority of tumor", 8522/3 identifies both invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma and is the most accurate description.

Rule H19 Code the **subtype/variant** (specific histology) **ONLY** when there is a NOS/NST and a subtype/variant **AND** the subtype/variant is documented to be **greater than 90%** of the tumor.

Note 1: When a histology is listed as "minimal", "focus/foci/focal", "microscopic", you can assume the other histological portion comprises greater than 90% of the tumor.

Note 2: Use **Table 3** to identify **NOS/NST** and subtypes/variants. Examples include the following:

- Carcinoma NST **8500** and a subtype/variant of carcinoma NST
- Glycogen-rich clear cell carcinoma **8315** and a subtype/variant of glycogen-rich clear cell carcinoma
- Lobular carcinoma **8520** and a subtype/variant of lobular carcinoma
- Medullary carcinoma **8510** and a subtype/variant of medullary carcinoma
- Metaplastic carcinoma **8575** and a subtype/variant of metaplastic carcinoma
- Papillary carcinoma **8503** and a subtype/variant of papillary carcinoma
- Sarcoma **8800** and a subtype/variant of sarcoma
- Small cell carcinoma **8041** and a subtype/variant of small cell carcinoma

Note 3: **Do not** code any histology described as **features or differentiation** unless it is part of the preferred term.

Example 1: Pathology from excision shows a 1.4 cm tumor and a diagnosis of clear cell carcinoma 8310/3 with a focus of glycogen-rich clear cell carcinoma NOS 8315/3. Because the glycogen-rich clear cell carcinoma NOS is just a focus, more than 90% of the tumor is clear cell carcinoma. Code the subtype/variant: clear cell carcinoma 8310/3.

Example 2: Pathology from an excised tumor says tumor is 95% metaplastic carcinoma squamous cell carcinoma 8070/3 and the remainder is metaplastic carcinoma NOS 8575/3. Code the subtype/variant: squamous cell carcinoma 8070/3.

Rule H20 Code the **NOS/NST** when there is a NOS/NST and a subtype/variant **AND**

- The subtype/variant is designated as **less than or equal to 90%** of tumor **OR**
- The percentage of each is **unknown/not documented**

Example 1: Pathology diagnosis is carcinoma NST 8500/3 and pleomorphic carcinoma 8022/3. The percentage of subtype/variant is unknown. Code the NOS: carcinoma NST 8500/3.

Example 2: Pathology says the majority of tumor is metaplastic carcinoma with chondroid differentiation 8571/3 and the remainder is metaplastic carcinoma NOS 8575/3. Majority simply means greater than 50%, so it is unknown whether or not the subtype/variant is greater than 90% of the tumor. Code metaplastic carcinoma NOS 8575/3.

Rule H21 Code the histology that comprises **greater than 50%** of tumor when **two histologies** are:

- On **different rows** in **Table 3** in the Equivalent Terms and definitions **OR**
- **Different subtypes** of the same NOS **OR**
- A **combination code** from **Table 2** and a code from **Table 3**

Note 1: This rule does not apply to mucinous. See previous rules.
Note 2: The rules are hierarchical, so the tumors are **NOT** a NOS/NST and subtype/variant.
Note 3: If the majority histology is unknown/not documented, continue through the rules.
Example: Pathology reads the tumor is predominantly carcinoma NST 8500/3 with areas of tubular carcinoma 8211/3. Code the predominant histology: carcinoma NST 8500/3. Carcinoma NST and tubular carcinoma are on different rows in Table 3, so they are distinctly different histologies.

Rule H22 Code a **combination code** when there are **two histologies** (two components) within a single tumor and the majority histology is unknown/not documented.

Note 1: Use **Table 2** in the Equivalent Terms and Definitions to identify valid combination codes.
Note 2: The rules are hierarchical, so the tumors are **NOT** a NOS/NST and a single subtype/variant.
Note 3: The diagnosis may be two subtypes/variants and the pathologist may mention the presence of duct/carcinoma NST. Ignore the mention of carcinoma NST.
Note 4: **Do not** use a combination code when the second histology is described as **features or differentiation** unless it is part of the preferred term.
Note 5: The histologies may be identified as:

- Mixed histologies
- Combination histologies
- Histology 1 **AND** histology 2
- Histology 1 **WITH** histology 2

This is the end of instructions for a Single Tumor: Invasive Only

Code the histology according to the rule that fits the case

Multiple Tumors Abstracted as a Single Primary

Note 1: DCIS is often multifocal/multicentric; use the Single Tumor: In Situ module.
Note 2: First use the multiple primary rules to ensure that the multiple tumors are to be abstracted as a single primary.

Rule H23 Code the **underlying tumor** when there is a diagnosis of inflammatory carcinoma[2]:

Example: The patient has a clinical diagnosis of inflammatory breast carcinoma. Pathology shows carcinoma NST with dermal invasion as well as erythema. Code the underlying tumor: carcinoma NST 8500/3.

Informational item: The **clinical symptoms** of inflammatory breast cancer include rapid breast enlargement and skin changes (redness, edema peau d'orange) involving more than a third of the breast. Usually there is a diffuse firmness of the breast and there is no palpable underlying mass.

Note 1: Record the inflammatory carcinoma in **staging** fields.
Note 2: Code inflammatory carcinoma 8530/3 when it is the only diagnosis available (DCO, outpatient only, no follow-up).

Rule H24 Code **Paget disease** and **ductal carcinoma** as follows when:

- Pathology specifies Paget disease as **invasive** /3 **OR** behavior not documented **AND**
- Underlying tumor is:
  - Invasive carcinoma NST/duct carcinoma **8541/3**
  - DCIS **8543/3**

Note: Ignore the presence of lobular carcinoma in situ (LCIS).

Rule H25 Code Paget disease and DCIS **8543/2** when there is Paget disease (specified as **in situ**) with underlying **DCIS**.

Rule H26 Code the histology when only **one histology** is present in **all** tumors.

Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in Table 3 and coding errors may occur if the table is not used.
Note 2: When the histology is not listed in **Table 3,** use the ICD-O and all updates.
Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 3, ICD-O or all updates.

Rule H27 Code the **invasive** histology when there are both invasive and in situ histologies.

Exception: Continue through the rules when there are multiple tumors of ductal and lobular carcinoma with different behaviors.

Rule H28 Code **8522** when carcinoma NST and lobular are present in multiple tumors.

- DCIS and in situ lobular **8522/2**
- DCIS and pleomorphic lobular carcinoma in situ **8522/2**
- Invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma (includes invasive pleomorphic lobular carcinoma) **8522/3**
- Intraductal and invasive lobular carcinoma (includes invasive pleomorphic lobular carcinoma) **8522/3**
- Infiltrating duct and lobular carcinoma in situ (LCIS) **8522/3**
- Infiltrating duct and pleomorphic lobular carcinoma in situ **8522/3**
- Infiltrating lobular carcinoma and ductal carcinoma in situ (DCIS) **8522/3**
- Infiltrating pleomorphic lobular carcinoma and ductal carcinoma in situ (DCIS) **8522/3**

Note 1: Assign behavior code /3 even when an in situ histology is mixed with an invasive. This aligns with ICD-O-3.2 and was vetted with specialty matter experts.

Note 2: CAP uses the term **Invasive carcinoma with ductal and lobular features** ("mixed type carcinoma") as a synonym for duct carcinoma/carcinoma NST AND lobular carcinoma 8522/3.

Note 3: One tumor may be carcinoma NST and the other lobular, or all tumors may be a mixture of carcinoma NST and lobular.

Note 4: This combination code specifically identifies carcinoma NST and lobular carcinoma. For all other histological combinations, continue through the rules.

Rule H29 Code the **NOS/NST** when there is a NOS/NST and a subtype/variant:

- Mixed in all of the tumors **OR**
- Separate tumors with different histologies

Note: It is very difficult to determine whether the subtype/variant is greater than 90% of the tumor mass when there are multiple tumors.

Breast Histology Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule H30 Code a **combination code** when there are **two histologies** (two components) within all tumors.

Note 1: Use **Table 2** in the Equivalent Terms and Definitions to identify valid combination codes.

Note 2: **Do not** use a combination code when the second histology is described as **differentiation or features,** unless it is part of the preferred term.

Note 3: The histologies may be identified as:

- Mixed histologies
- Combination histology
- Histology 1 **AND** histology 2
- Histology 1 **WITH** histology 2

Note 4: Table 2 is used for **two** histologies. When there are **greater than two** histologies, use the "last resort" code **8255** because none of the other combinations include greater than two histologies.

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

Code the histology according to the rule that fits the case

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Introduction

Note 1: New terms and codes in these rules are based on the ***WHO Classification of Tumors of the Digestive System*** 2010 edition.

Note 2: Ninety-eight percent of colon cancers are **adenocarcinoma** and adenocarcinoma **subtypes.**

Note 3: **Mixed histologies and specific variants or subtypes of adenocarcinoma** other than mucinous/colloid or signet ring cell are **rare**. A less common combination is **mixed adenoneuroendocrine carcinoma** (**MANEC**) **8244** (previously called adenocarcinoma and carcinoid). The new terminology was originally proposed for tumors arising from goblet cell carcinoid but with more aggressive adenocarcinoma histology. It was also proposed because **carcinoids** are a subgroup of **neuroendocrine carcinoma**. Pathologists **may still diagnose** adenocarcinoma and carcinoid, adenocarcinoid, or adenocarcinoma and a specific neuroendocrine tumor or adenocarcinoma arising from/with a NET (including specific types of **NET-like goblet cell** carcinoid). Over time, the histologic diagnoses will change to MANEC.

Note 4: **De novo (previously called frank) adenocarcinoma** arises in the mucosa of the bowel, not in a polyp.

Note 5: **Terms Seen More Frequently: NET, NEC, GIST**

- **NET** (neuroendocrine tumor): The term NET is gradually replacing **carcinoid;** however, some pathologists still use the term carcinoid
- **NEC** (neuroendocrine carcinoma): The term NEC includes **small cell neuroendocrine** carcinoma, **large cell neuroendocrine** carcinoma, and **poorly differentiated neuroendocrine** carcinoma
- **GIST** (gastrointestinal stromal tumor):
  - GISTs were originally thought to be smooth muscle tumors but are now thought to originate from the interstitial cells of Cajal, neuro-regulatory cells in the GI tract. Prior to the implementation of an ICD-O-3 histology code for GISTs in **2001**, they were reported as a GI sarcoma, usually **leiomyosarcoma**
  - GISTs are more common in the stomach (60%) and small intestine (30%), but 1-2% occur in the colon and 3% in the rectum
  - About a quarter of gastric GISTs are **malignant**
  - It is often difficult for the pathologist to determine the **behavior** of a GIST
  - GIST NOS becomes a reportable neoplasm beginning with cases diagnosed 1/1/2021 forward

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

Note 7: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2017: Use 2007 MPH Rules

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

Note 8: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Changes from 2007 MPH Rules

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

1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, 2018 Rules instruct "Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor)."
2. **Rectum** and **Rectosigmoid** are now included with the Colon Rules. In the 2007 MPH Rules, they were included with Other Sites.
3. There are new multiple primary rules which address **anastomotic recurrence.**
4. Neuroendocrine tumors (formerly carcinoid) arising in the appendix are reportable for cases diagnosed 1/1/2015 and forward.
5. **Rule clarification: Pseudomyxoma peritonei** (accumulation of mucin-secreting tumor cells in the abdominal or pelvic cavity) now has a **two-tiered system** (WHO 2010) that classifies pseudomyxoma peritonei as either **high-grade** or **low-grade** (see below)**.** Pseudomyxoma peritonei is usually associated with **mucinous** tumors of the appendix and is rarely associated with ovarian mucinous tumors.
    - **High-**grade pseudomyxoma peritonei is **malignant** /3
    - **Low**-grade pseudomyxoma peritonei is **not malignant** /1
    - See **Histology Rules** for **coding instructions**

6. There are **dysplasias** which have been assigned an **in situ behavior** code /2 in **WHO** and in **the ICD-O Update**. Despite becoming a /2, they are **not reportable in the US**. They are reportable in Canada.
    - Dysplasia **was not** collected in the past. If dysplasia is added to the database with the same code as in situ tumors, there will be a **huge upsurge** in the **incidence** of in situ neoplasms. The various agencies are looking for solutions to this issue.
    - There would be no way to **separate** the dysplasias from the in-situ neoplasms in the database, which would cause problems with surveillance (long-term studies) since the prognosis and probabilities of disease progression are different between an in-situ tumor and a dysplasia
    - **Pathologists frequently use the term "severe dysplasia" or "high grade dysplasia" in place of carcinoma in situ. Code CIS only** if the pathologist expressly states "CIS"
7. **Polyps** are now **disregarded** when coding histology. For example, adenocarcinoma in an adenomatous polyp is coded as adenocarcinoma 8140. For the purposes of determining multiple primaries, tumors coded as adenocarcinoma in a polyp for pre-2018 cases should be treated as adenocarcinoma 8140.

New codes/terms are identified by asterisks (*) in the histology table in the Terms and Definitions.

New for 2022

1. Timing changes to rules M7 and M8: The timing for subsequent tumors at the anastomosis has changed from 24 months to 36 months. The change is effective for cases diagnosed beginning 1/1/2022 forward. For cases diagnosed 1/1/2018 through 12/31/2021, the timing rule remains at 24 months.
2. Low grade appendiceal neoplasm (LAMN) will become reportable effective for cases diagnosed 1/1/2022 forward. LAMN may be either in situ 8480/2 or malignant 8480/3 based on physician statement of behavior. LAMN diagnosed prior to 1/1/2022 are not reportable.

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Carcinoid; NET; neuroendocrine tumor
- Carcinoma; adenocarcinoma;
  - A histology type must be stated for these terms to be equal
  - **Example**: Mucinous carcinoma and mucinous adenocarcinoma are both coded 8480
- De novo; frank adenocarcinoma (obsolete)
- Familial polyposis; familial adenomatous polyposis (FAP) **8220**
- Intramucosal; lateral extension within the mucosal layer of the GI tract
- Invasion through colon wall; extension through colon wall; transmural
  Note: The term "**transmural**" is used to describe **extension through all layers** of the wall, but not past the wall **OR extension through the serosa into the mesentery**. **Read** the **pathology** report carefully.
- Mucinous; mucoid; mucous; colloid
- Neuroendocrine carcinoma; NEC
- Polyp; adenoma; polyp NOS; adenomatous polyp
  Note 1**:** The term "**polyp**" means projecting from a surface.
  Note 2**:** There are many kinds of polyps. Most common are **adenomas**, which are part of the adenoma-cancer sequence.
  Note 3**: Other types o**f polyps include hyperplastic, juvenile, Peutz-Jeghers and serrated adenoma/polyp.
- Serosa; visceral peritoneum
- Simultaneous; existing at the same time; concurrent; prior to first course treatment
- Site; topography
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a standard manner in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician's statement** that the term is malignant/cancer
  - These terms are used **ONLY** to determine multiple primaries
  - **Do not** use these terms for casefinding or determining reportability
- Type; subtype; variant

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Terms that are NOT Equivalent or Equal

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

- Carcinoma, NOS 8010 is not equivalent to adenocarcinoma, NOS 8140
- **Component** is not equivalent to **subtype/type/variant**
  Note: Component is only coded when the pathologist specifies the component as a second **carcinoma.**
- **Exophytic** and **polypoid** are not equivalent to either an **adenoma** or an **adenomatous polyp**. The terms "exophytic" and "polypoid" refer to anything projecting from the bowel mucosa into the lumen. The lesion may be benign, malignant, or inflammatory.
- **Phenotype** is not equivalent to **subtype/type/variant**
- **Polypoid adenocarcinoma** is not equivalent to **adenocarcinoma in a polyp**

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Table 1: Specific Histologies, NOS, and Subtypes/Variants

Use Table 1 as directed by the **Histology Rules** to assign the more common histology codes for malignancies found in the colon, rectosigmoid and rectum.

Note 1: Rare histologies may not be listed in the table. When a histology term is not found, reference ICD-O and all updates.
Note 2: Submit a question to Ask a SEER Registrar when the histology code is not found in Table 1, ICD-O or all updates.
Note 3: Behavior codes are listed when the term has only one possible behavior (either a /2 or /3). For histologies which may be either /2 or /3, a behavior code is not listed. Code behavior from pathology.

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

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

Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
Adenocarcinoma 8140 ***Note 1*: See Histology Rules** for instructions on coding adenocarcinoma **subtypes/variants** arising in a polyp ***Note 2*:** When the term **intestinal adenocarcinoma** is used to describe a colon primary, it simply means the	Adenocarcinoma, NOS Adenocarcinoma/carcinoma in a polyp NOS (now coded to 8140) Adenocarcinoma/carcinoma in adenomatous polyp (now coded to 8140) Adenocarcinoma/carcinoma in polypoid adenoma (now coded to 8140) Adenocarcinoma/carcinoma in serrated adenoma (now coded to 8140) Adenocarcinoma and mucinous carcinoma, mucinous documented as	Adenoid cystic carcinoma **8200** Cribriform comedo-type carcinoma/ adenocarcinoma, cribriform comedo-type **8201*** Diffuse adenocarcinoma/carcinoma **8145** High grade appendiceal mucinous neoplasm (HAMN)/Low grade appendiceal mucinous neoplasm **8480- see Note 3** Linitis plastica **8142/3** Medullary adenocarcinoma/carcinoma **8510** Micropapillary carcinoma **8265*** Mucinous/colloid adenocarcinoma/carcinoma **8480** Mucoepidermoid carcinoma **8430** Serrated adenocarcinoma **8213***

Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
appearance is similar to adenocarcinoma seen in the stomach and is coded to adenocarcinoma NOS **8140** **Note 3:** Effective 1/1/2022, LAMN becomes reportable and is coded 8480/2, unless the pathologist indicates invasive behavior, which is coded 8480/3. HAMN can be either /2 or /3 depending on the pathologist statement of behavior.	less than 50% of tumor OR percentage of mucinous unknown/not documented Adenocarcinoma and signet ring cell carcinoma, percentage of signet ring cell carcinoma documented as less than 50% of tumor OR unknown/not documented Adenocarcinoma/carcinoma in tubular polyp (now coded to 8140) Adenocarcinoma/carcinoma in tubulovillous polyp (now coded to 8140) Adenocarcinoma/carcinoma in villous adenoma (now coded to 8140) Adenocarcinoma in any type of polyp Adenocarcinoma, intestinal type Adenocarcinoma and cribriform carcinoma percentage of cribriform documented as less than 50% of tumor OR unknown/not documented Adenocarcinoma with mucinous and signet ring cell features Comedocarcinoma Intestinal adenocarcinoma	Signet ring cell/poorly cohesive adenocarcinoma/carcinoma **8490** Superficial spreading adenocarcinoma **8143** Tubulopapillary carcinoma **8263** Undifferentiated adenocarcinoma/carcinoma **8020**
Adenosquamous carcinoma 8560 ***Note:*** This code **cannot be used** for **adenocarcinoma subtypes/variants** with squamous cell/epidermoid carcinoma	Mixed adenocarcinoma NOS and epidermoid carcinoma Mixed adenocarcinoma NOS and squamous cell carcinoma	

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Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
Combined small cell carcinoma 8045	Small cell carcinoma mixed with • Adenocarcinoma **OR** • Neuroendocrine carcinoma **OR** • Any other type of carcinoma/adenocarcinoma	
Gastrinoma 8153		
Gastrointestinal stromal tumor 8936/3 ***Note*:** See standard setter reportability guidelines.	Gastrointestinal autonomic nerve tumor GANT Gastrointestinal pacemaker cell tumor Gastrointestinal stromal tumor GIST, NOS GIST, malignant GIST, spindle cell type Gastrointestinal stromal sarcoma Succinate dehydrogenase-deficient gastrointestinal stromal tumor	
Mixed adenoneuroendocrine carcinoma 8244	Adenocarcinoma ex-goblet cell Adenocarcinoma mixed with high-grade large cell neuroendocrine carcinoma Adenocarcinoma mixed with high-grade small cell neuroendocrine carcinoma MANEC Mixed neuroendocrine carcinoma	Goblet cell adenocarcinoma/Goblet cell carcinoid **8243**
Mixed neuroendocrine non-neuroendocrine neoplasm 8154	MiNEN	
Neuroendocrine carcinoma 8246	NEC	Large cell NEC **8013** Small cell NEC **8041**

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Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
Neuroendocrine tumor Grade 1 (G1) 8240 ***Note*: When the diagnosis is exactly "carcinoid"** it may be a Grade 1 or Grade 2 NET. Default is coding NET Grade 1 **8240**.	Carcinoid NOS Low-grade neuroendocrine tumor NET Grade 1 (G1) Well differentiated neuroendocrine tumor	EC cell serotonin-producing NET/enterochromaffin cell carcinoid **8241** Neuroendocrine tumor (NET) Grade 2 (G2) **8249** Somatostatin-producing NET **8156**
Sarcoma NOS 8800/3		Angiosarcoma/hemangiosarcoma **9120/3** Leiomyosarcoma **8890/3**
Spindle cell carcinoma 8032		
Squamous cell carcinoma 8070	Epidermoid carcinoma NOS Squamous cell carcinoma NOS Squamous cell epithelioma	

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

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 2: Histologies Not Reportable for Colon, Rectosigmoid and Rectum

Column 1 lists the **non-reportable** histology term and code for NOS or specific
Column 2 lists the **synonym**(s) for the term
Column 3 lists the **subtype/variant** of the NOS term with the histology code
Column 4 lists the **reason** these histologies are **not reportable**

Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
Adenoma 8140/0 ***Note*:** No malignancy in polyps	Adenoma NOS	Tubular adenoma **8211/0** Tubulovillous adenoma **8263/0** Villous adenoma **8261/0**	Non-malignant
Adenomatous polyp, high grade dysplasia 8210/2			Non-reportable terminology
Cowden-associated polyp No code ***Note*:** No malignancy in polyps	Cowden disease Cowden syndrome Multiple hamartoma syndrome		Non-malignant /no code
Dysplasia, high grade 8148/2 ***Note:*** Colorectal primaries only (C180-C189, C199 and C209)	High-grade dysplasia Intraepithelial neoplasia, high grade		**CURRENTLY NOT REPORTABLE**
Dysplasia, low grade 8148/0* ***Note:*** Colorectal primaries only (C180-C189, C199 and C209)	Intraepithelial neoplasia, low grade		Non-malignant

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Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
Familial adenomatous polyposis (FAP) No code	Adenomatous polyposis coli Bussey-Garder polyposis Familial multiple polyposis Familiar polyposis coli Familial polyposis of the colon and rectum Familial polyposis of the gastrointestinal tract Gardner syndrome Multiple adenomatosis		Reportable only when there is cancer in a polyp
Gangliocytic paraganglioma 8683/0			Non-malignant
Gastrointestinal stromal tumor stated as benign, borderline, or non-malignant 8936/1 (SEE NOTE in column 2)	GIST NOS GIST, behavior not specified ***Note***: Gastrointestinal stromal tumor, NOS is not reportable for cases diagnosed prior to 1/1/2021. Cases diagnosed 1/1/2021 forward are reportable		Non-malignant
Hyperplastic polyp No code			Non-malignant/no code
Inflammatory or pseudopolyp No code			Reactive lesions; mimic carcinoma
Intestinal-type adenoma, high grade 8144/2			Non-reportable terminology

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Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
Juvenile polyp No code	Combined juvenile polyposis/hereditary Hemorrhagic telangietasis (Osler-Webec-Rendu) syndrome Familial juvenile polyposis Generalized juvenile polyposis Hamartomatous gastrointestinal polyposis; Juvenile polyposis Juvenile polyposis coli Juvenile polyposis of infancy		Non-malignant / no code
L cell glucagon-like peptide and PP/PYY-producing NETs 8152/1*			Non-malignant
Leiomyoma 8890/0			Non-malignant
Lipoma 8850/0			Benign accumulation of fat cells that are circumscribed or encapsulated
Low-grade appendiceal mucinous neoplasm 8480/1 ***Note*:** May have low-grade, non-invasive pseudomyxoma peritonei, mucinous implants in peritoneum or beyond	LAMN		Non-malignant **Note:** LAMN is non-reportable for cases diagnosed **prior to 1/1/2022**. Beginning 1/1/2022, LAMN becomes a reportable neoplasm- See Table 1
Lynch syndrome No code			Non-malignant/no code

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
Mesenchymal tumors		Granular cell tumor **9580/0** Hemangioma **9120/0**	Non-malignant
Peutz-Jeghers polyp No code	Intraepithelial neoplasia in Peutz-Jeghers polyp(s) Periorificial lentiginosis Peutz-Jeghers polyposis Polyps-and-spots syndrome		Non-malignant/no code
Pseudomyxoma peritonei (when pathologist does not designate as malignant OR implants are benign) **8480/1**			Non-malignant. When both implants and site of origin are benign, the case is not reportable.
Serrated dysplasia, high grade 8213/2			Non-reportable terminology
Sessile serrated adenoma/polyp 8213/0* ***Note:*** No malignancy in polyps	Serrated polyposis Sporadic serrated polyps Traditional serrated adenoma		Non-malignant
Tubular adenoma, high grade 8211/2			Non-reportable terminology
Tubular carcinoid, no malignancy 8245/1			Non-malignant
Tubulovillous adenoma, high grade 8263/2			Non-reportable terminology
Villous adenoma, high grade 8261/2			Non-reportable terminology

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

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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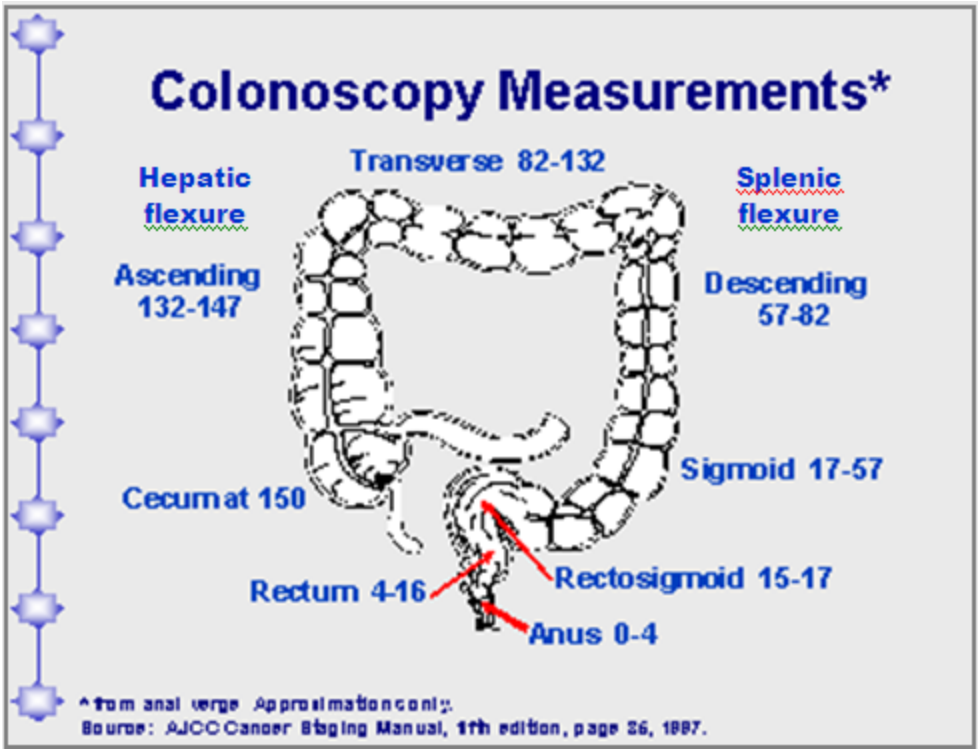
Illustrations

Colonoscopy measurements which may be used to determine primary site when no site is designated



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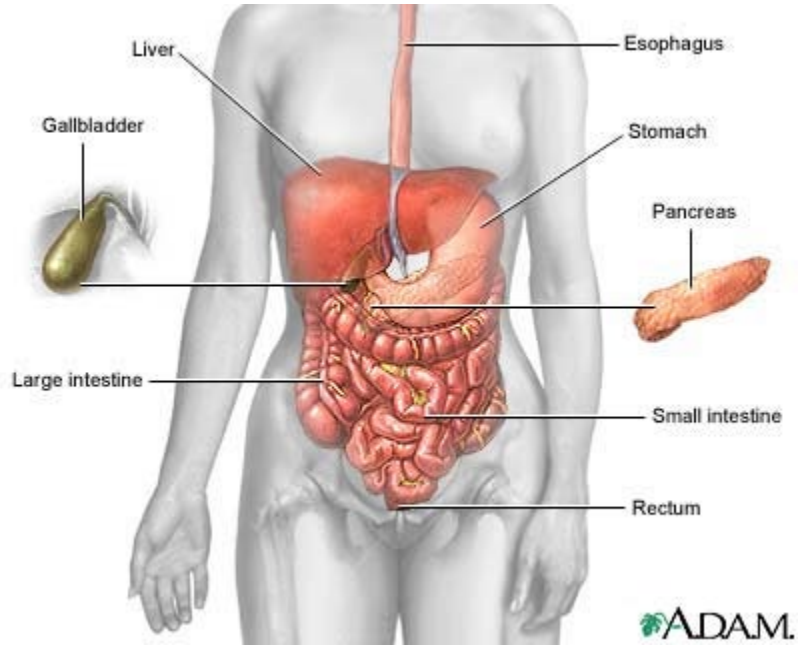
GI System



Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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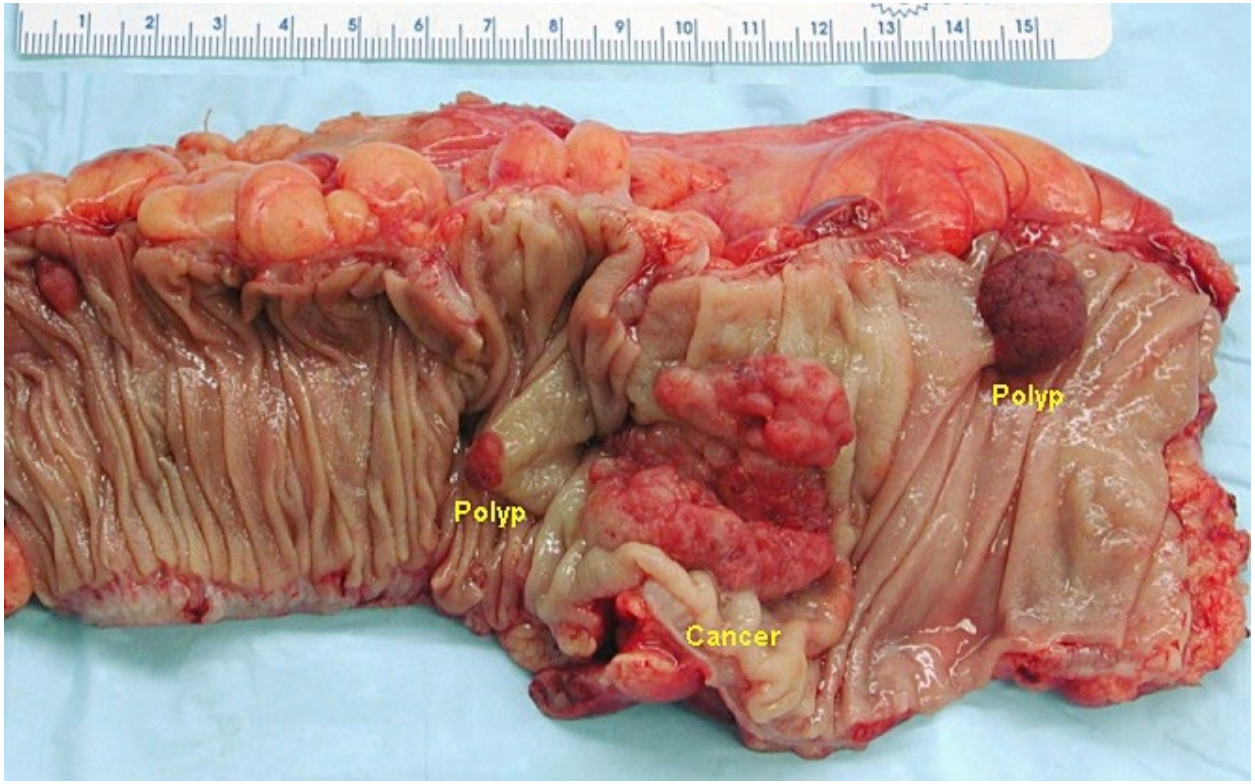
Polyps and de novo or "frank" adenocarcinoma in colon



Source: http://upload.wikimedia.org/wikipedia/commons/4/44/Colon_cancer.jpg

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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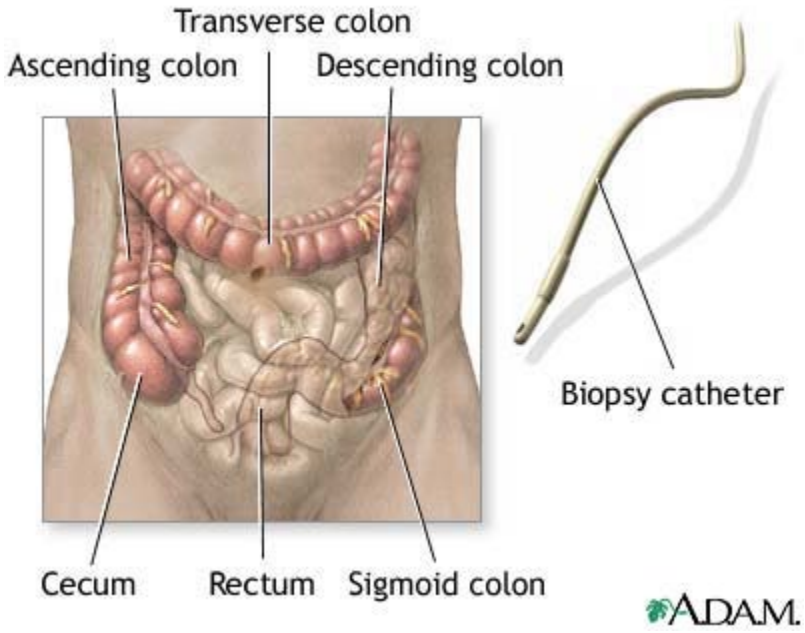
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

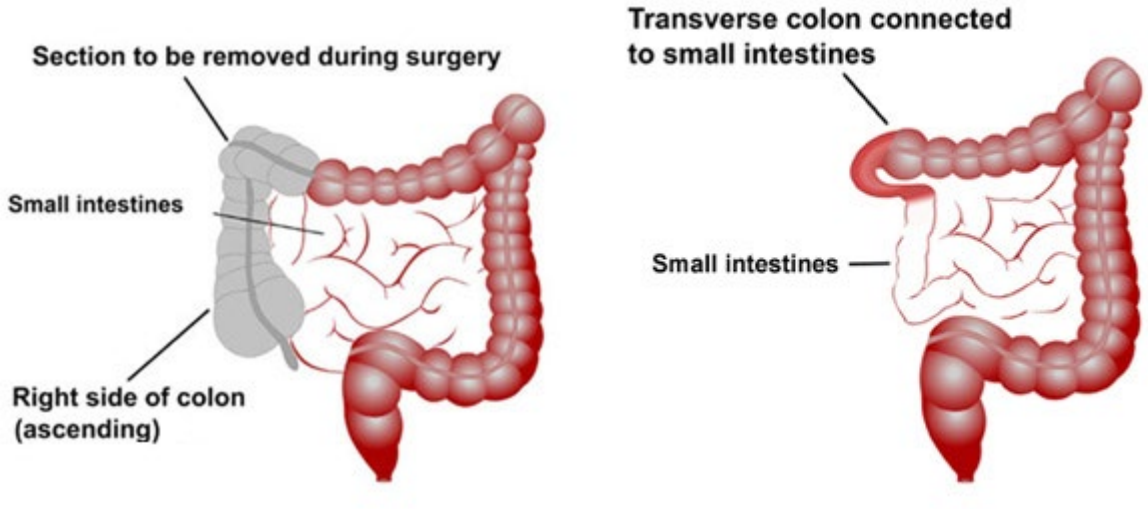
Large intestine; snare instrument to remove polyps



Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Colon Surgery: Hemicolectomy

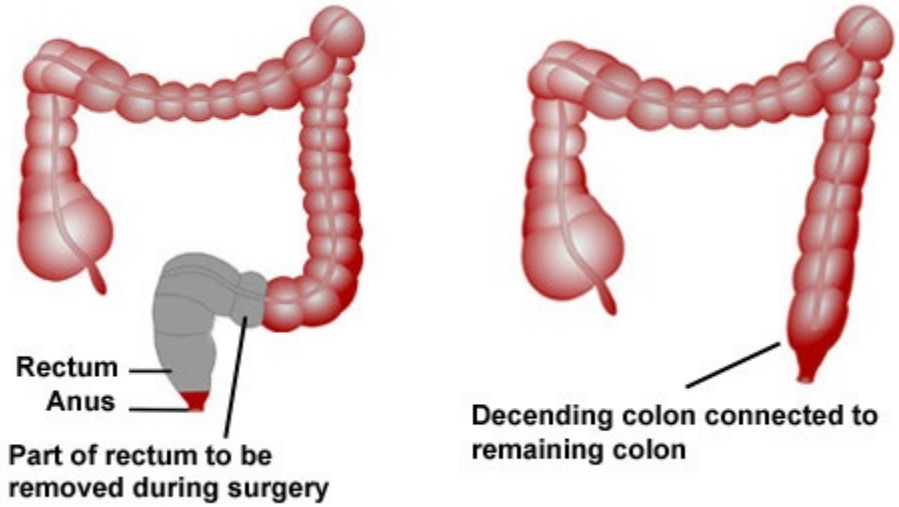
The primary treatment for colon cancer is surgery. Part of the large bowel and the surrounding lymph nodes are removed. The remaining bowel is then joined together (anastomosis).



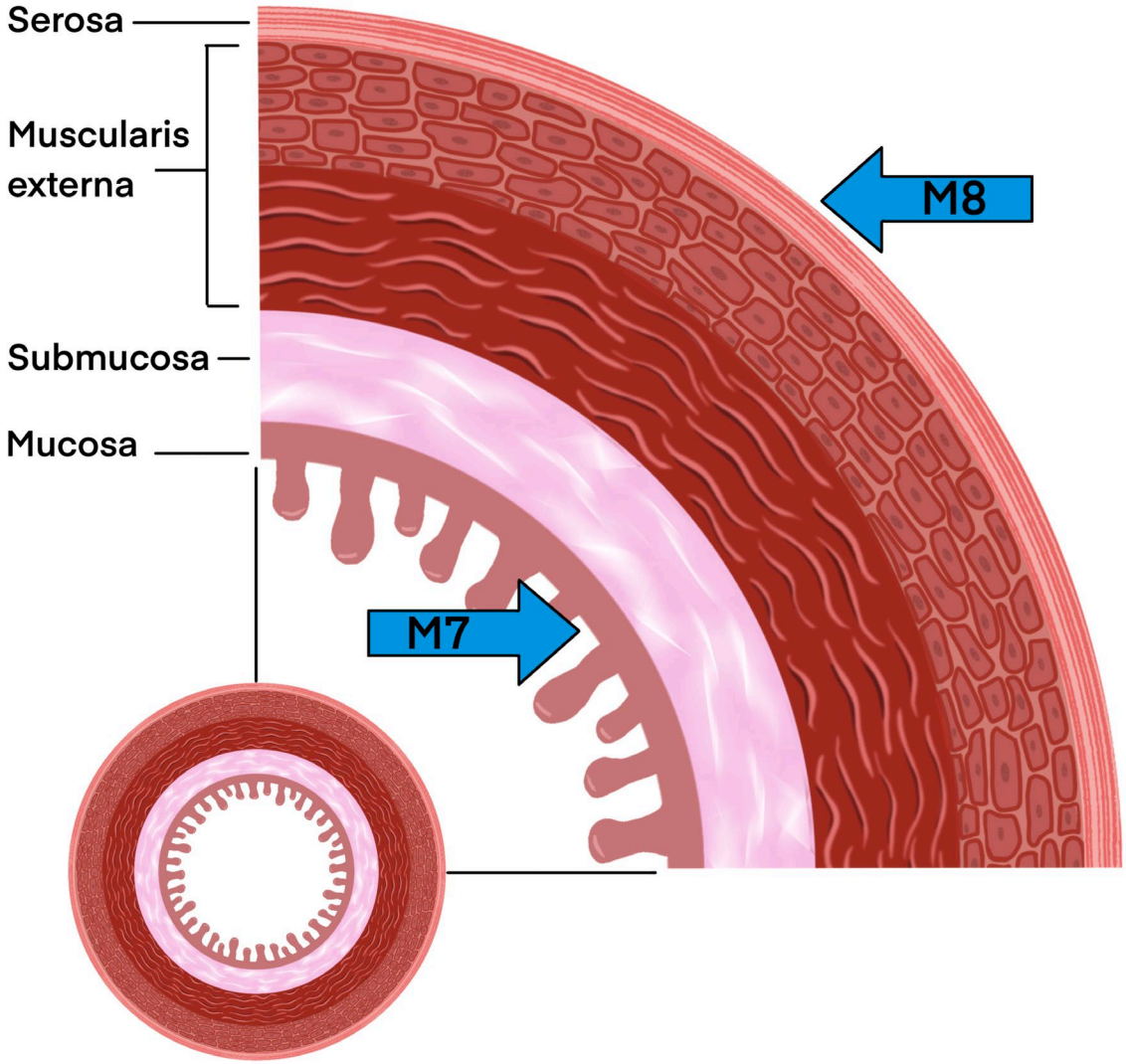
http://www.cedars-sinai.edu/Patients/Programs-and-Services/Colorectal-Cancer-Center/Services-and-Treatments/Rectal-Cancer.aspx

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rectal Surgery



Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Discontinuous lesions in soft tissue adjacent to primary site
- Regional or distant lymph nodes for the primary site being abstracted as identified in Summary Staging Manual
- Brain
- Liver
- Lung
- Peritoneum
- Spinal cord (not frequent)

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown if Single or Multiple Tumors

Note: **Collision tumors** are counted as **two individual** tumors for the purpose of determining multiple primaries. Collision tumors were originally two **separate** tumors that arose in close proximity. As the tumors increased in size, they merged or overlapped each other. Use the Multiple Tumors module.

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors

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

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Note: **Collision tumors** are counted as **two individual** tumors for the purpose of determining multiple primaries. Collision tumors were originally two **separate** tumors that arose in close proximity. As the tumors increased in size, they merged or overlapped each other. Use the Multiple Tumors module.

Rule M2 Abstract a **single primary**[i] when there is a **single tumor.**
Note 1: A single tumor is always a single primary.
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor

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

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multiple Tumors

Note 1: Multiple tumors may be a single primary or multiple primaries.
Note 2: **Collision tumors** are counted as **two individual** tumors for the purpose of determining multiple primaries. Collision tumors were originally two **separate** tumors that arose in close proximity. As the tumors increased in size, they merged or overlapped each other. Use the Multiple Tumors module to determine if they are a single primary or multiple primaries.

Rule M3 Abstract a **single primary**[i] when there is adenocarcinoma in situ and/or invasive in at least one polyp **AND**

- There is a clinical diagnosis of familial polyposis (FAP) **OR**
- Greater than 100 polyps are documented (no diagnosis of FAP)

Note 1: A diagnosis of familial polyposis (FAP) is made when the patient has **greater than 100** adenomatous polyps. Polyps with adenocarcinoma and benign polyps will be present. Because there are many polyps, the pathologist does not examine every polyp.

Note 2: **In situ /2** and **malignant /3** adenocarcinoma in polyps, malignancies with remnants of a polyp, as well as de novo (previously called frank) malignancies may be present in **multiple segments** of the colon or in both the **colon** and **rectum**. Polyposis **may** be present in other GI sites such as stomach (a de novo does not have to be present; all adenocarcinoma may be in polyps).

Note 3: FAP is a **genetic** disease. The characteristics of FAP are **numerous precancerous polyps** in the colon and rectum when the patient reaches puberty. If not treated, the polyps typically become malignant. Patients often have **total colectomies.**

Note 4: **Multiple polyps** in the colorectum is **not equivalent** to FAP.

Note 5: Code **primary site** as follows:

- Present in more than one segment of colon: **C189** colon, NOS
- Present in colon and rectosigmoid **OR** colon and rectum: **C199** rectosigmoid junction
- Present in colon and small intestine: **C260** intestinal tract, NOS (there is no code for large and small bowel)

  Note: In addition to the colon and small intestine, FAP may also be present in the:
  - Stomach **AND/OR**
  - Rectosigmoid **AND/OR**
  - Rectum

Example: The patient has a diagnosis of FAP. The operative report and physician's documentation say that polyps with adenocarcinoma were present in specimens removed from the ascending colon and the sigmoid colon. The ascending and sigmoid colon are part of the large bowel. Code the primary site **C189** colon NOS.

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M4 Abstract **multiple primaries**[ii] when there are separate, non-contiguous tumors in sites with ICD-O site codes that **differ** at the second C**X**xx and/or third Cx**X**x character.

Note 1: Definition of separate/non-contiguous tumors: at least two malignancies which **do not** overlap/merge.

Note 2: **Differences** at either the **second** or **third** characters are different primary sites/multiple primaries.

Example 1: Breast C**50**x and colon C**18**x

Example 2: Colon C**18**x and rectum C**20**9 (This does not include FAP- see earlier rules)

Note: This rule **does not** apply to a single **overlapping** malignancy of colon and rectum.

Rule M5 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more different **subtypes/variants** in Column 3, **Table 1** in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS**: Medullary carcinoma NOS 8510/3 and tubulopapillary adenocarcinoma 8263/3 are both subtypes of adenocarcinoma NOS 8140/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Goblet cell carcinoid 8243/3 is a subtype of mixed adenoneuroendocrine carcinoma 8244/3; somatostatin-producing NET 8156/3 is a subtype of neuroendocrine tumor Grade 1 (G1) 8240/3. They are distinctly different histologies. Abstract multiple primaries.

Rule M6 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are on **different rows in Table 1** in the Equivalent Terms and Definitions. Timing is irrelevant.

Note: Each row in the table is a **distinctly different** histology.

Rule M7 Abstract **multiple primaries**[ii] when a subsequent tumor arises at the **anastomotic** site **AND:**

- One tumor is a **NOS** and the other is a **subtype/variant** of that NOS **OR**
- The subsequent tumor occurs **greater than 36 months** after original tumor resection **OR**
  Note: For cases diagnosed prior to 1/1/2022, the time interval is greater than 24 months.
- The **subsequent** tumor arises in the **mucosa** (see illustration)
  Note: Bullet three does not apply to GIST. GISTs only start in the wall; never in the mucosa.

Example: (For bullet 1: NOS and subtype/variant) The original tumor was adenocarcinoma NOS **8140**. The patient had a hemicolectomy. There was a recurrence at the **anastomotic** site diagnosed exactly as **mucinous** adenocarcinoma **8480.** Mucinous adenocarcinoma is a subtype/variant of the NOS adenocarcinoma, but they are two different histologies. **Code two primaries,** one for the original adenocarcinoma NOS and another for the subsequent anastomotic site mucinous adenocarcinoma.

Note 1: There may or may not be **physician documentation** of anastomotic recurrence. Follow the rules.

Note 2: When the original tumor was diagnosed prior to 1/1/2018 and was coded to adenocarcinoma in a polyp, and the anastomotic site tumor is adenocarcinoma per 2018 rules, the tumors are the same histology. ICD-O codes differ because of changes in histology coding rules.
Note 3: The tumor may or may not invade into the colon wall or adjacent tissue.
Note 4: A "rectal stump" is an anastomotic site.
Note 5: These rules are hierarchical. Only use this rule when previous rules do not apply.

Rule M8 Abstract a **single primary**[i] when a subsequent tumor arises at the **anastomotic** site **AND:**

- The subsequent tumor occurs **less than or equal to 36 months** after original tumor resection **OR**
  Note: For cases diagnosed prior to 1/1/2022, timing is less than or equal to 24 months
- The tumor arises in **colon/rectal wall** and/or surrounding tissue; there is **no involvement** of the **mucosa** (see illustration) **OR**
- The pathologist or clinician documents an **anastomotic recurrence**

Note 1: Bullet two does not apply to GIST. GISTs only start in the wall; never in the mucosa.
Note 2: The physician may stage the subsequent tumor because the depth of invasion determines the second course of treatment.
Note 3: These tumors are a single primary/**recurrence**. Registrars that collect recurrence information should record the information in the recurrence fields.
Note 4: A "rectal stump" is an anastomotic site.

Rule M9 Abstract **multiple primaries**[ii] when there are separate, non-contiguous tumors in sites with ICD-O site codes that **differ** at the fourth characters C18**X**.

Note: Differences at the fourth character include different segments of the colon. Abstract a primary for each separate non-contiguous tumor in a different segment of the colon. This rule is not used for colon NOS C189. C189 is rarely used other than DCO.

Example: The patient has adenocarcinoma in situ in a **sigmoid** polyp and mucinous adenocarcinoma in a polyp in the **descending** colon, the site code differs at the fourth character (sigmoid C18**7** and descending C18**6**). **Code two primaries**, one for the sigmoid and another for the descending colon.

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M10 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being **clinically disease-free** for **greater than one year** after the original diagnosis or last recurrence.

Note 1: **Clinically** disease-free means that there was **no evidence** of recurrence on follow-up.
- Colonoscopies are NED
- Scans are NED

Note 2: When there is a recurrence less than or equal to one year of diagnosis, the **"clock"** starts over. The time interval is calculated from the **date of last recurrence.** In other words, the patient must have been **disease-free** for **greater than one year** from the date of the last recurrence.

Note 3: When the first course of treatment was a **polypectomy** only, this rule means there were **no recurrences** for greater than one year.

Note 4: When the first course of treatment was a **colectomy or A&P resection**, there were **no anastomotic recurrences** for greater than one year.

Note 5: When it is **unknown/not documented** whether the patient had a recurrence, default to **date of diagnosis** to compute the time interval.

Note 6: The physician may state this is a **recurrence,** meaning the patient had a previous colon tumor and now has another colon tumor. **Follow the rules;** do not attempt to interpret the physician's statement.

Rule M11 Abstract a **single primary**[i] when synchronous, separate/non-contiguous tumors are on **the same row in Table 1** in the Equivalent Terms and Definitions.

Note: The same row means the tumors are:
- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3)

Rule M12 Abstract a **single primary**[i] (the invasive) when an **in situ** tumor is diagnosed **after** an **invasive** tumor.

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.

Note 2: The tumors **may** be a **NOS** and a **subtype/variant** of that NOS. See **Table 1** in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

Note 3: The **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M13 Abstract a **single primary**[i] (the invasive) when an invasive tumor is diagnosed **less than or equal to 60 days** after an **in situ** tumor.

Note 1: The rules are hierarchical**.** Only use this rule when previous rules do not apply.

Note 2: **Change behavior** code on original abstract from /2 to /3. **Do not** change **date of diagnosis**.

Note 3: If the case has already been submitted to the central registry, **report** all changes.

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

Note 5: See the **COC** and **SEER manuals** for **instructions** on coding **data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M14 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60 days** after an **in situ** tumor.

Note 1: Abstract both the invasive and in situ tumors.

Note 2: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression. This rule is based on **long-term epidemiologic** studies of **recurrence intervals.** The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were authors, co-authors, or editors of the AJCC Staging Manual.

Rule M15 Abstract a **single primary**[i] when tumors do not meet any of the above criteria.

Note: Use caution when applying this default rule. Please confirm that you have not overlooked an applicable rule.

Example: The pathology states adenocarcinoma in situ 8140/2 and a second non-contiguous invasive adenocarcinoma 8140/3 in the sigmoid colon C187. Multiple tumors that are the same histology in the same primary site (same four characters of ICD-O topography code) are a single primary.

This is the end of instructions for Multiple Tumors.

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[i] Prepare one abstract. Use the histology rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

Colon, Rectosigmoid, and Rectum Histology Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

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

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

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

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

This is a hierarchical list of source documentation.
Code the **most specific** pathology/tissue from either **resection** or **biopsy**.

Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

1. **Tissue or pathology report from primary site** (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol

   Note 1: Addendums and comments on the pathology report are given a high priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
   Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.
   Note 3: The CAP protocol is a checklist which:

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

2. Tissue/pathology from a **metastatic** site
   Note 1: Code the behavior /3.
   Note 2: The tissue from a metastatic site often shows **variations** from the primary tumor. When it is the only tissue available, it is more accurate than a scan.
3. **Scan:** The following list is in **priority order**.
   A. CT
   B. PET
   C. MRI
4. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following **priority order:**
   A. Treatment plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that **refers to original pathology, cytology, or scan(s)**
   D. Physician's **reference** to type of cancer (**histology**) in the medical record
   Note 1: Code the specific histology when documented.
   Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented.
5. **Cytology** (seldom used for colon, rectosigmoid and rectum)

Colon, Rectosigmoid, and Rectum Histology Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Coding Histology

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

1. **Code** the **most specific** histology or **subtype/type/variant, regardless of whether it is described as:**
   A. The majority or predominant part of tumor
   B. The minority of tumor
   C. A component
   Note 1: Mucinous and signet ring cell carcinoma must meet a percentage requirement in order to be coded. Refer to the Histology Rules if mucinous and/or signet ring cell carcinoma is present.
   Note 2: The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.
   Example: When the diagnosis is adenocarcinoma with a component of medullary **carcinoma**, code medullary carcinoma 8510.
   Negative Example: When the diagnosis is simply adenocarcinoma with a medullary component, code adenocarcinoma NOS 8140. Do not assume this is a medullary carcinoma. This could be medullary differentiation or features.
   Note 3: When the most specific histology is described as differentiation or features, see #2.
   Example 1: Diagnosis for a single tumor is adenocarcinoma 8140 with the majority or predominant part of tumor being medullary adenocarcinoma 8510. Code the subtype/variant: medullary adenocarcinoma 8510.
   Example 2: Diagnosis for a single tumor is mixed neuroendocrine carcinoma 8244 with minority of tumor being goblet cell carcinoid 8243. Code the subtype/variant: goblet cell carcinoid 8243.
   Example 3: Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.

2. Code the histology 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".
   Note: Do not code differentiation or features when there is no specific ICD-O code.

3. Code the specific histology described by **ambiguous terminology** (list follows) **ONLY** when A or B is true:
   A. The only diagnosis available is **one histology** term described by ambiguous terminology
      - CoC and SEER require reporting of cases diagnosed only by ambiguous terminology

- Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

Example: Outpatient biopsy says probably malignant GIST. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology malignant GIST. The case meets the criteria in #**3A.**

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

Example 1: The pathology diagnosis is adenocarcinoma consistent with micropapillary carcinoma. The oncology consult says the patient has micropapillary carcinoma of the cecum. This is clinical confirmation of the diagnosis, code micropapillary carcinoma. The case meets the criteria in **bullet 1.**

Example 2: The pathology diagnosis is mixed neuroendocrine carcinoma consistent with goblet cell carcinoid. The treatment plan says the patient will receive the following treatment for goblet cell carcinoid. Treatment plan confirms goblet cell carcinoid; code goblet cell carcinoid. The case meets the criteria in **bullet 2.**

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

List of Ambiguous Terminology

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

4. **Do not code** histology when described as:

    - Architecture
    - Foci; focus; focal
    - Pattern

Colon, Rectosigmoid, and Rectum Histology Rules
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule H1 Code adenocarcinoma with neuroendocrine differentiation **8574** when the final diagnosis is **exactly** "adenocarcinoma with neuroendocrine differentiation".

Note: **Do not** use this code when:

- The diagnosis is any subtype/variant of adenocarcinoma with neuroendocrine differentiation
- Any modifier other than differentiation is used, i.e., adenocarcinoma with neuroendocrine features

Rule H2 Code the **histology** and **ignore the polyp** when a carcinoma **originates** in a **polyp.**

Note 1: This is a **change** from the **2007** MPH rules which instructed registrars to use the codes for malignancies in a polyp, such as adenocarcinoma in a polyp **8210.**

Note 2: Sufficient data has been collected to:

- Determine the frequency with which carcinomas arise within polyps
- Establish patient care guidelines for individuals with colon polyps

Example: Colonoscopy with polypectomy finds mucinous adenocarcinoma in the polyp. Code mucinous adenocarcinoma **8480.**

Rule H3 Code combined small cell carcinoma **8045** when the final diagnosis is **small cell** carcinoma **AND any other carcinoma.**

Examples:

- Small cell carcinoma **8041** and adenocarcinoma **8140**
- Small cell carcinoma **8041** and neuroendocrine carcinoma **8246**

Rule H4 Code **mixed mucinous and signet ring cell** as follows:

- Adenocarcinoma with mucinous and signet ring features – code adenocarcinoma **8140**
- Mucinous carcinoma and signet ring cell carcinoma:
  - Mucinous carcinoma documented as **greater than 50%** – code mucinous carcinoma **8480**
  - Signet ring cell carcinoma documented as **greater than 50%** – code signet ring cell carcinoma **8490**
  - Percentage of mucinous carcinoma and signet ring cell carcinoma **unknown/not designated**- code adenocarcinoma mixed subtypes **8255**

Note: This rule is for mucinous carcinoma and signet ring cell carcinoma in a single tumor. For mucinous adenocarcinoma mixed with another histology OR signet ring cell carcinoma mixed with another histology, proceed through the rules.

Rule H5 Code **low grade appendiceal mucinous neoplasm (LAMN)** and **high grade appendiceal mucinous neoplasm (HAMN) 8480/2** when:

- Diagnosis date is 1/1/2022 forward **AND**
- Behavior is stated to be in situ/non-invasive **OR**
- Behavior is not indicated

Note 1: ICD-O-3.2 lists LAMN with behavior of /1. WHO 5th Ed Digestive Systems Tumors indicates this neoplasm is considered in situ. After consulting with WHO Digestive System editors, College of American pathologists, and AJCC GI chapter experts, the standard setting organizations have agreed LAMN should be collected and should be assigned a behavior code of /2 beginning with cases diagnosed 1/1/2022 forward.

Note 2: A diagnosis of LAMN or HAMN does not require the tumor be comprised of greater than 50% mucinous in order to be coded 8480.

Note 3: If the pathologist indicates LAMN or HAMN is invasive or has a malignant behavior, continue through the rules.

Rule H6 Code invasive **mucinous** adenocarcinoma **8480** when the diagnosis is any of the following:

- **Exactly** "**mucinous** adenocarcinoma" (no modifiers)
- High grade appendiceal mucinous neoplasm (HAMN) stated to be invasive (DX 1/1/2022 forward)
- **High-grade** pseudomyxoma peritonei
- **Invasive** pseudomyxoma peritonei
- Low grade appendiceal mucinous neoplasm (LAMN) stated to be invasive (DX 1/1/2022 forward)
- **Malignant** pseudomyxoma peritonei
- Two histologies and mucinous is documented to be **greater than 50%** of the tumor
  - Mucinous carcinoma must meet a percentage requirement in order to be coded. Do not use majority of tumor, predominantly, or predominant part of the tumor to code mucinous 8480.

Note 1: Be very **careful** when **determining primary** site; almost all pseudomyxoma peritonei originate in the appendix C181. However, it **can be metastatic** disease from sites such as bowel, ovary, or bladder. Code the primary site as designated by a physician. When the primary site is not designated, code unknown primary **C809** and the histology as mucinous carcinoma **8480.**

Note 2: Report the appendiceal mucinous neoplasm as malignant /3 using the ICD-O matrix principle and the SEER and COC Manuals when the **pathology** from the appendix is **low-grade mucinous** neoplasm (not reportable prior to 1/1/2022) **AND**

- The pseudomyxoma peritonei are **high-grade**/invasive/malignant **OR**
- Patient is **treated** for malignant pseudomyxoma peritonei OR
- The diagnosis is low grade appendiceal mucinous neoplasm (LAMN) and the physician states it is malignant OR

- The diagnosis is high grade appendiceal mucinous neoplasm (HAMN) and the physician states it is malignant
- The pathologist has staged the LAMN as T3 or T4

Note 3: The following are **non-reportable for cases diagnosed prior to 1/1/2022:**

- Appendiceal neoplasm with **low-grade** pseudomyxoma peritonei **AND no treatment**
- **No designation** of high- or low-grade for the appendiceal neoplasm **AND no treatment** for the pseudomyxoma peritonei

Rule H7 Code invasive **signet ring cell adenocarcinoma 8490** when the diagnosis is any of the following:

- Exactly **signet ring cell carcinoma** (no modifiers)
- **Adenocarcinoma and signet ring cell carcinoma**, where signet ring cell is documented to be **greater than 50%** of the tumor
  - Signet ring cell adenocarcinoma must meet a percentage requirement in order to be coded. Do not use majority of tumor, predominantly, or predominant part of tumor to code signet ring cell 8490.

Rule H8 Code adenocarcinoma NOS **8140** when the final diagnosis is:

- Two histologies:
  - Adenocarcinoma and mucinous carcinoma
    - Percentage of mucinous **unknown/not documented**
    - Mucinous documented as less than or equal to 50% of tumor
  - Adenocarcinoma and signet ring cell carcinoma
    - Percentage of signet ring **unknown/not documented**
    - Signet ring cell documented as less than or equal to 50% of tumor
- **Exactly** adenocarcinoma **OR**
- **Intestinal** type adenocarcinoma **OR** adenocarcinoma intestinal type (no modifiers or additional histologic terms).

Note 1: Code **8140 adenocarcinoma NOS** even if pathology says intestinal type adenocarcinoma.

Note 2: Do **not** use code **8144** adenocarcinoma intestinal type for **colorectal** primaries. Intestinal type adenocarcinoma 8144 is used for tumors which occur in the stomach, head and neck, and specific GYN sites. It is called intestinal because it resembles carcinoma which occurs in the colon, rectosigmoid or rectum.

Note 3: When a diagnosis of intestinal type adenocarcinoma is **further described** by a **specific term** (such as mucinous intestinal type adenocarcinoma or signet ring cell intestinal type adenocarcinoma), it would be treated as an adenocarcinoma with a **subtype/variant**.

Rule H9 Code the histology when only **one histology** is present.

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

Note 2: Use the ICD-O and all updates when the histology is not listed in Table 1.

Note 3: Submit a question to Ask a SEER Registrar when the histology code is not found in Table 1, ICD-O or all updates.

Rule H10 Code the **invasive** histology **when in situ** and **invasive** histologies are present in the **same tumo**r.

Rule H11 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Mixed adenoneuroendocrine carcinoma **8244** and a subtype/variant of mixed adenoneuroendocrine carcinoma
- Neuroendocrine carcinoma **8246** and a subtype/variant of neuroendocrine carcinoma
- Neuroendocrine tumor Grade 1 (G1) **8240** and a subtype/variant of neuroendocrine tumor Grade 1 (G1)
- Sarcoma **8800** and a subtype/variant of sarcoma

Note 1: See **Table 1** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

Note 2: Only code subtypes/variant when pathology gives an **exact diagnosis**. **Do not** code the subtype/variant when **modified** by terms such as **differentiation, features of, etc., unless** there is a specific code for the histology term with the modifier.

This is the end of instructions for Single Tumor.

Code the histology using the rule that fits the case.

Colon, Rectosigmoid, and Rectum Histology Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multiple Tumors Abstracted as a Single Primary

Note: Multiple tumors **must be a single primary** to use this module. **See** the **Multiple Primary Rules** to determine whether these tumors are a single primary.

Rule H12 Code adenocarcinoma in familial adenomatous polyposis coli (**FAP**) **8220** when **clinical** history says the patient has **familial polyposis AND**

- The final diagnosis on the **pathology report** from resection is **adenocarcinoma in FAP OR**
- There are **greater than 100 polyps** identified in the resected specimen

Note 1: Use this rule **only** when there are **multiple polyps**. **Do not** use for a single polyp (adenoma) or for a de novo (frank) malignancy and a malignancy in a single polyp.
Note 2: Use this rule **ONLY** for adenocarcinoma in **FAP.**
Note 3: The disease process, treatment, and prognosis for FAP is not as favorable as a single polyp with adenocarcinoma.

Rule H13 Code adenocarcinoma in multiple adenomatous polyps **8221** when FAP is not mentioned **AND**

- There are at least 2 polyps with adenocarcinoma /2 or /3 **AND**
  - Less than or equal to 100 polyps are identified **OR**
  - The exact number of polyps is unknown/not documented

Note 1: **Do not use** this code for a malignancy in **a single polyp** (adenoma) or for a de novo (frank) malignancy.
Note 2: Use this rule **ONLY** for **adenocarcinoma NOS** in multiple polyps**.**

Rule H14 Code the histology of the **invasive** tumor when there are **in situ** /2 and **invasive** /3 tumors.

- One tumor is in situ and the other is invasive
- All tumors are a **mixture** of **in situ** and **invasive** histology

Rule H15 Code the histology when only **one** histology is present in **all** tumors.

Note 1: Use **Table 1** to code histology. New codes, terms, and synonyms are included in **Table 1** and coding errors may occur if the table is not used.
Note 2: When the histology is **not listed** in **Table 1,** use the **ICD-O** and all **updates**.
Note 3: Submit a question to Ask a SEER Registrar when the histology code is not found in Table 1, ICD-O or all updates.

Rule H16 Code the **subtype/variant** when the diagnosis is a **NOS** and a **single subtype/variant** of that NOS such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Mixed adenoneuroendocrine carcinoma **8244** and a subtype/variant of mixed adenoneuroendocrine carcinoma
- Neuroendocrine carcinoma **8246** and a subtype/variant of neuroendocrine carcinoma
- Neuroendocrine tumor Grade 1 (G1) **8240** and a subtype/variant of neuroendocrine tumor Grade 1 (G1)
- Sarcoma **8800** and a subtype/variant of sarcoma

Note 1: All tumors may be **mixed** histologies (NOS and a subtype/variant of that NOS) **OR** one tumor may be a **NOS** histology and the other tumor a **subtype/variant** of that NOS.

Note 2: See **Table 1** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

Note 3: Check the **Multiple Primary Rules** to confirm that the tumors are a single primary.

Note 4: Only code subtypes/variant when pathology gives an **exact diagnosis**. **Do not** code the subtype/variant when **modified** by terms such as **differentiation, features of, etc., unless** there is a specific code for the histology term with the modifier.

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

Code the histology using the rule that fits the case.

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Introduction

Note 1: Tables and rules refer to ICD-O rather than ICD-O-3. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Note 3: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Changes from 2007 MPH Rules

1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, 2018 Rules instruct "Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor)."
2. Two bone sites, mandible C411 and maxilla C410, have been added to the Head and Neck Rules.
3. Autonomic nervous system C479 has been added as a primary site for those paragangliomas reported as malignant.

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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

New for 2022

1. The 2018 Solid Tumor Head and Neck Rules, Table 5, instruct squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086) are coded only when HPV status is determined by tests based on ISH, PCR, RT-PCR technologies to detect the viral DNA or RNA. P16 was not a valid test to assign these codes. **Beginning with cases diagnosed 1/1/2022 forward, p16 test results can be used to code squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086).**
2. Beginning 1/1/2022, non-keratinizing squamous cell carcinoma, HPV positive is coded 8085 for sites listed in Table 5 only. A diagnosis of non-keratinizing squamous cell carcinoma, NOS is coded 8072.
3. Beginning 1/1/2022, keratinizing squamous cell carcinoma, HPV negative is coded 8086 for sites listed in Table 5 only. A diagnosis of keratinizing squamous cell carcinoma, NOS is coded 8071.

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Equivalent or Equal Terms

These terms can be used interchangeably:

- Adenocarcinoma; adenocarcinoma NOS; carcinoma; carcinoma NOS
- And; with
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**
- Contiguous; continuous
- Hemangiosarcoma; angiosarcoma
- Hypopharynx; laryngopharynx
- In situ; noninvasive; intraepithelial
- Malignant tumor; malignant mass; malignant lesion; malignant neoplasm
- Simultaneous; existing at the same time; concurrent; prior to first course treatment
- Site; topography
- Squamous cell carcinoma; squamous carcinoma; squamous cell epithelioma; epidermoid carcinoma
- Squamous cell carcinoma, HPV-negative; squamous cell carcinoma, HPV-independent (8086)
- Squamous cell carcinoma, HPV-positive; squamous cell carcinoma, HPV-associated; squamous cell carcinoma, HPV-related (8085)
- Squamous cell carcinoma with verrucous **growth** pattern; squamous cell carcinoma
  - Growth pattern is not a histological type
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms **unless** there is a **physician's statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to **determine** multiple **primaries**
  - **Do not** use these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

Head and Neck Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Terms that are NOT Equivalent or Equal

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

- **Component** is not equivalent to **subtype/type/variant**
  Note: Component is **only** coded when the pathologist specifies the component as a second **carcinoma**
- **p16 positive** is not equivalent to **HPV positive (pre-2022)**
- **p16 negative** is not equivalent to **HPV negative (pre-2022)**
- **Phenotype** is not equivalent to **subtype/type/variant**
- **Squamous cell carcinoma with prominent keratinization 8070** is not equivalent to **keratinizing squamous cell carcinoma 8071**
- Salivary **gland** adenocarcinoma **8140** is not equivalent to salivary **duct** carcinoma **8500**

Coding Primary Site When There is Conflicting Information

Identifying the primary site is **difficult** because:

- Workups (PE scans, endoscopies, biopsies) each provide a unique view of the tumor, therefore the medical record often contains conflicting documentation on the primary site.
- The sites/organs are small and right next to each other. Tumors frequently extend into adjacent anatomic sites, or overlap multiple contiguous sites.

Priority Order for Identifying Primary Site When There is Conflicting Information

Note: Record primary site based on the most definitive indication of primary site in the medical documentation and use the priority order when there is conflicting info without a definitive statement.

1. **Tumor Board**
   A. Specialty
   B. General

2. **Tissue/pathology** from tumor resection or biopsy
   A. Operative report
   B. Addendum and/or comments on tissue/pathology report
   C. Final diagnosis on issue/pathology report
   D. CAP protocol/summary

3. **Scans**
   A. CT
   B. MRI
   C. PET

4. **Physician documentation**. Use the documentation in the following priority order:
   A. Physician's **reference** in medical record to primary site from **original pathology, cytology, or scan(s), any other documentation**
   B. Physician's **reference** to primary site in the medical record

5. Use **Tables 1-9** to assist in assigning primary site when a **SINGLE** lesion overlaps two or more sites.
   A. Go to the appropriate table for each involved site (use the hyperlinked index below).
   B. Compare the histology diagnosis to the histologies in the table for each of the involved sites.
   C. When the histology diagnosis is listed for only one primary site (only listed in one table), code that primary site.

6. When the primary site cannot be determined using previous instructions, code as follows for an overlapping lesion:
   A. **C028** Overlapping lesion of tongue (See **Table 4** for subsites of the tongue)
   B. **C058** Overlapping lesion of palate, junction of hard and soft palate (See **Table 4** for subsites of the palate)
   C. **C088** Overlapping lesion of major salivary glands (See **Table 6** for specific salivary glands)
   D. **C148** Overlapping lesion of lip, oral cavity and pharynx
   Note: Codes and terms for overlapping lesions C__.8 are **not** included in the **tables**

7. Code to the NOS region
   A. **C069** Mouth NOS (See **Table 4** for mouth subsites)
   B. **C089** Major Salivary Gland NOS (See **Table 6** for specific salivary glands)
   C. **C099** Tonsil NOS (See **Table 5** for tonsil subsites)
   D. **C109** Oropharynx NOS (See **Table 5** for oropharynx subsites)
   E. **C119** Nasopharynx NOS (See **Table 2** for nasopharynx subsites)

F. **C139** Hypopharynx NOS (See **Table 3** for hypopharynx subsites)
G. **C140** Pharynx NOS

Note: Pharynx NOS includes the oropharynx, nasopharynx, and hypopharynx.

H. **C760** Head, face, or neck NOS (organs involved unknown/not documented)

Note: This code is used in circumstances such as biopsy of lymph node and no information about primary site

- Patient lost to follow-up; no further information available
- Patient/family declined further work-up or treatment

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Table Index

Head and Neck Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 1: Tumors of Nasal Cavity, Paranasal Sinuses and Skull base

Table 1 lists the more common histologies for the following head and neck subsites:

- **C300** Nasal cavity; naris; nasal cartilage; nasal mucosa; nasal septum NOS; nasal turbinate; nostril; vestibule of nose
- **C310** Maxillary sinus; maxillary antrum; antrum NOS
- **C311** Ethmoid sinus
- **C312** Frontal sinus
- **C313** Sphenoid sinus
- **C318** Overlapping lesion of accessory sinuses
- **C319** Accessory sinus NOS; accessory nasal sinus; paranasal sinus

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.
Note: Hematopoietic tumors are common to the nasal cavity and paranasal sinuses.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including rhabdomyosarcoma 8900/3 (column 3). Rhabdomyosarcoma is also a NOS because it has subtypes/variants. The subtypes/variants are indented under the NOS (rhabdomyosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that rhabdomyosarcoma has subtypes/variants.

When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, rhabdomyosarcoma **8900/3** and Alveolar rhabdomyosarcoma **8920/3** are a NOS and a subtype/variant, **NOT** two different subtypes.

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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenocarcinoma 8140 ***Note:*** Adenocarcinoma intestinal-type of the sinonasal tract is morphologically similar to adenocarcinomas of the intestines	Adenocarcinoma non-intestinal type Low-grade adenocarcinoma Renal cell-like carcinoma Seromucinous adenocarcinoma TAC Terminal tubulous adenocarcinoma Tubulopapillary low-grade adenocarcinoma	Adenocarcinoma intestinal type (ITAC) **8144** Colloid-type adenocarcinoma **8144** Colonic-type adenocarcinoma **8144** Enteric-type adenocarcinoma **8144**
Lymphoepithelial carcinoma 8082	LEC Lymphoepithelioma-like carcinoma	
Malignant peripheral nerve sheath tumor 9540/3	MPNST Neurofibrosarcoma	Malignant neurilemoma/malignant schwannoma **9560/3**
Mucoepidermoid carcinoma 8430	Salivary gland-type mucoepidermoid carcinoma	
Mucosal melanoma 8720		
Myoepithelial carcinoma 8982	Myoepithelioma, malignant	
NUT carcinoma 8023*	Midline carcinoma of children and young adults with NUT rearrangement NUT midline carcinoma	
Olfactory neuroblastoma 9522/3	Esthesioneuroblastoma Olfactory placode tumor ONB	Esthesioneurocytoma 9521/3 Esthesioneuroepithelioma/Olfactory neuroepithelioma 9523/3

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Primitive neuroectodermal tumor 9364	Adult neuroblastoma Ewings sarcoma Peripheral neuroblastoma Peripheral neuroectodermal tumor Peripheral neuroepithelioma	
Sarcoma 8800/3 ***Note 1:*** Angiosarcomas are coded to the organ in which they occur. The prognosis and disease process of angiosarcomas differ between sites Contiguous organs, blood vessels, and lymph nodes are not the same for every organ. ***Note 2:*** Rhabdomyosarcoma **8900/3** has subtypes/variants: Alveolar rhabdomyosarcoma **8920/3** Embryonal rhabdomyosarcoma **8910/3** Pleomorphic rhabdomyosarcoma, adult type **8901/3**		Angiosarcoma/hemangiosarcoma **9120/3** Biphenotypic sinonasal sarcoma (BSNS)/low-grade sinonasal sarcoma with neural and myogenic features **9045/3*** Epithelioid hemangioendothelioma **9133/3** Fibrosarcoma/adult-type fibrosarcoma **8810/3** Leiomyosarcoma **8890/3** Malignant hemangioendothelioma **9130/3** Rhabdomyosarcoma **8900/3** Alveolar rhabdomyosarcoma **8920/3** Embryonal rhabdomyosarcoma **8910/3** Pleomorphic rhabdomyosarcoma, adult type **8901/3** Spindle cell rhabdomyosarcoma **8912/3** Synovial sarcoma/synovial cell sarcoma **9040/3** Undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma **8802/3**
Sinonasal undifferentiated carcinoma 8020 ***Note:*** This is an undifferentiated carcinoma of the Sinonasal tract.	Sinonasal carcinoma, undifferentiated SNUC	

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Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Squamous cell carcinoma 8070 ***Note:*** Sinonasal squamous cell tumors account for about 3% of head and neck malignancies.	Squamous cell carcinoma, usual type **8070/3** Conventional Squamous cell carcinoma NOS Epidermoid carcinoma, NOS **8070/3** Epidermoid carcinoma in situ, NOS **8070/2** Squamous carcinoma **8070/3** Squamous cell carcinoma in situ, NOS **8070/2** Squamous cell epithelioma **8070/3** Intraepithelial squamous cell carcinoma **8070/2**	Basaloid squamous cell carcinoma **8083** Keratinizing squamous cell carcinoma (KSCC) **8071** Epidermoid carcinoma, keratinizing Squamous cell carcinoma, large cell, keratinizing Papillary squamous cell carcinoma **8052** Squamous cell carcinoma, large cell, nonkeratinizing/Squamous cell carcinoma, nonkeratinizing, NOS **8072** Schneiderian carcinoma/cylindrical cell carcinoma **8121** Sarcomatoid squamous cell carcinoma/spindle cell squamous cell carcinoma (SC-SCC) **8074** Verrucous carcinoma **8051**
Teratocarcinosarcoma 9081	Blastoma Malignant teratoma Teratocarcinoma Teratoid carcinosarcoma	

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

Table 2: Tumors of Nasopharynx

Table 2 lists the more common histologies for the following head and neck subsites:

- **C110** Superior wall of nasopharynx; roof of nasopharynx
- **C111** Posterior wall of nasopharynx only (does not include adenoid/pharyngeal tonsil)
- **C112** Lateral wall of nasopharynx; fossa of Rosenmuller
- **C113** Anterior wall of nasopharynx; nasopharyngeal surface of soft palate; pharyngeal fornix; choana; posterior margin of nasal septum
- **C118** Overlapping lesion of nasopharynx. Use only when a **single lesion** overlaps **subsites** of the nasopharynx.
  Example: A single tumor overlaps C110 superior wall of nasopharynx and C111 posterior wall of the nasopharynx.
- **C119** Nasopharynx NOS; nasopharyngeal wall; use when a specific subsite cannot be identified.
  Example: The primary site is designated as pharyngeal wall. It is unknown whether it is the superior, posterior lateral, or anterior wall.

Note 1: The **nasopharynx** is the upper part of the pharynx. It is above the soft palate and extends to the nasal passages.
Note 2: Nasopharyngeal tumors are usually assigned to the subsite in which they occur.

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.
Note: Hematopoietic tumors are common to the nasopharynx.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

Table begins on next page.

Head and Neck Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenoid cystic carcinoma 8200		
Chordoma 9370		
Nasopharyngeal papillary adenocarcinoma 8260	Thyroid-like low-grade nasopharyngeal; papillary adenocarcinoma	
Squamous cell carcinoma NOS 8070		Basaloid squamous cell carcinoma **8083** Keratinizing squamous cell carcinoma **8071** Non-keratinizing squamous cell carcinoma **8072** Lymphoepithelial carcinoma **8082** Undifferentiated carcinoma/Undifferentiated carcinoma with lymphoid stroma **8020**

Table 3: Tumors of Pyriform Sinus, Hypopharynx, Larynx, Trachea, and Parapharyngeal Space

Table 3 lists the more common histologies for the following head and neck subsites:

- **C129** Pyriform sinus
- **C130** Postcricoid region; cricopharynx cricoid NOS
- **C131** Hypopharyngeal aspect of aryepiglottic fold; aryepiglottic fold NOS; arytenoid fold
- **C132** Posterior wall of hypopharynx
- **C138** Overlapping lesion of hypopharynx. Use only when a **single lesion overlaps subsites** of the hypopharynx.
  Example: A single tumor overlaps C130 postcricoid region and C131 aryepiglottic fold.
- **C139** Hypopharynx NOS and parapharyngeal space. Use only when the subsite/site is unknown
- **C320** Glottis; intrinsic larynx; laryngeal commissure; vocal cord NOS; true vocal cord; true cord
- **C321** Supraglottis; epiglottis NOS (excludes anterior surface of epiglottis); extrinsic larynx; laryngeal aspect of aryepiglottic fold; posterior surface of epiglottis; ventricular band of larynx; false vocal cord; false cord
- **C322** Subglottis
- **C323** Laryngeal cartilage; arytenoid cartilage; cricoid cartilage; cuneiform cartilage; thyroid cartilage
- **C328** Overlapping lesion of larynx
- **C329** Larynx NOS
- **C339** Trachea

Note 1: The **hypopharynx** is in the inferior position of the three segments of pharynx. The hypopharynx links the oropharynx to the esophagus, lower part of the pharynx. The pyriform sinus is located in the hypopharynx.
Note 2: The **larynx** is only 1 1/2 inches. It is inferior to the hyoid bone and tongue. It is anterior to the esophagus.
Note 3: The **trachea** starts where larynx ends and continues down the middle of the neck anterior to the esophagus.
Note 4: The **parapharyngeal space** is an equivalent of the lateral pharyngeal space which includes the soft tissue, vessels and skeletal muscles supporting the mechanics of the pharynx. Code the specific site when the soft tissue, vessel, or skeletal muscle is documented. When specific information is not available/not documented, code hypopharynx NOS, C139.
Note 5: These primary sites are mostly composed of muscle and cartilage, but the most common tumors arise from the epithelial lining of the structures (squamous cell carcinoma, for example).

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.
Note: Hematopoietic tumors are common to the hypopharynx, larynx and trachea.

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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenoid cystic carcinoma 8200	ACC (rare)	
Chondrosarcoma 9220/3	Chondrosarcoma grade 2/3 Chondrosarcoma NOS	Chondrosarcoma, grade 1 **9222/3** (cases diagnosed 1/1/2022 forward)
Liposarcoma 8850/3		Liposarcoma, well differentiated **8851/3**
Neuroendocrine tumor, NOS 8240/3	Carcinoid Neuroendocrine carcinoma grade 1 Well-differentiated neuroendocrine carcinoma	Large cell neuroendocrine carcinoma/LCNEC **8013/3** Neuroendocrine carcinoma grade 2/moderately-differentiated neuroendocrine carcinoma/atypical carcinoid **8249/3** Small cell neuroendocrine carcinoma/small cell carcinoma/SmCC **8041/3**
Squamous cell carcinoma (SCC) 8070	Epidermoid carcinoma Conventional Squamous cell carcinoma NOS	Adenosquamous carcinoma (ASC) **8560** Basaloid squamous cell carcinoma (BSCC) **8083** Lymphoepithelial carcinoma (LEC)/lymphoepithelioma-like carcinoma **8082** Keratinizing squamous cell carcinoma **8071** Non-keratinizing squamous cell carcinoma **8072** Papillary squamous cell carcinoma (PSCC) **8052** Spindle cell squamous cell carcinoma (SC-SCC) **8074** Verrucous squamous cell carcinoma (VC) **8051**

Table 4: Tumors of Oral Cavity and Mobile Tongue

Table 4 lists the more common histologies for the following head and neck subsites:

The **oral cavity category** includes the following:

Mobile Tongue:
- **C020** Dorsal surface of tongue NOS
- **C021** Border of tongue
- **C022** Ventral surface of tongue NOS
- **C023** Anterior 2/3 of tongue NOS
- **C028** Overlapping lesion of tongue
- **C029** Tongue NOS

Gum:
- **C030** Upper gum, maxillary gingiva, upper alveolar mucosa, upper alveolar ridge mucosa, upper alveolus, upper gingiva
- **C031** Lower gum mandibular gingiva, lower alveolar mucosa, lower alveolar ridge mucosa, lower alveolus, lower gingiva
- **C039** Gum NOS, gingiva NOS, alveolar mucosa NOS, alveolar ridge mucosa NOS, alveolar NOS periodontal tissue, tooth socket

Floor of Mouth:
- **C040** Anterior floor of mouth
- **C041** Lateral floor of mouth
- **C048** Overlapping lesion floor of mouth
- **C049** Floor of mouth NOS

Palate:
- **C050** Hard palate
- **C051** Soft palate
- **C052** Uvula
- **C058** Overlapping lesion of palate, junction of hard and soft palate
- **C059** Palate NOS, roof of mouth

Other and unspecified parts of Mouth:
- **C060** Cheek mucosa, buccal mucosa, internal cheek
- **C061** Vestibule of mouth, alveolar sulcus, buccal sulcus, labial sulcus

C062 Retromolar area, retromolar triangle, retromolar trigone
C068 Overlapping lesion of other and unspecified parts of mouth
C069 Mouth NOS, buccal cavity, oral cavity, oral mucosa, minor salivary gland NOS
Note: There is no ICD-O site code for minor salivary glands. Many minor salivary glands are located in the lips, inner cheek (buccal mucosa) and there are extensive minor salivary glands in the linings of the mouth and throat. Code to the site in which the salivary gland is located.

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.
Note: Hematopoietic tumors are common to the oral cavity.

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

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

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Mucoepidermoid carcinoma 8430	Mucoepidermoid tumor	
Myofibroblastic sarcoma 8825	Myofibrosarcoma	
Oral mucosal melanoma 8720		
Squamous cell carcinoma 8070	Conventional Squamous cell carcinoma NOS Squamous carcinoma Squamous cell carcinoma NOS	Acantholytic squamous cell carcinoma **8075** Keratinizing squamous cell carcinoma **8071** Non-keratinizing squamous cell carcinoma **8072** Verrucous squamous cell carcinoma 8051

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

Table 5: Tumors of the Oropharynx, Base of Tongue, Tonsils, Adenoids

Table 5 lists the more common histologies for the following head and neck subsites and coding histologies for cases diagnosed 1/1/2022 forward:

Cases diagnosed 1/1/2018 to 12/31/2021:
Squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086) are coded only when HPV status is determined by tests based on ISH, PCR, RT-PCR technologies to detect the viral DNA or RNA. p16 is not a valid test to assign these codes.

Cases diagnosed 1/1/2022 forward:
Beginning with cases diagnosed 1/1/2022 forward, p16 test results can be used to code squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086).

Cases diagnosed 1/1/2023 forward:
When the diagnosis is a subtype/variant of squamous cell carcinoma and HPV status is also noted, code the subtype/variant.
EXCEPTION: Keratinizing SCC (see Notes 1 and 2 in Table 5, Squamous Cell Carcinoma row)

- Example: Basaloid squamous cell carcinoma, HPV positive. Code basaloid SCC, 8083/3.

Oropharynx:

C100 Vallecula
C101 Anterior surface of epiglottis
C102 Lateral wall of oropharynx; lateral wall of nasopharynx
C103 Posterior wall of oropharynx; posterior wall of nasopharynx
C104 Brachial cleft
C108 Overlapping lesion of oropharynx; junctional region of oropharynx
C109 Oropharynx NOS; mesopharynx NOS; fauces NOS. Use this code only when the subsite has not been identified a subsite as the origin of the lesion.

Note: Code overlapping lesion of oropharynx; junctional region of oropharynx **C108** when a single tumor overlaps subsites of the oropharynx. For example, a single lesion which overlaps the vallecular and the anterior surface of the epiglottis.

C019 Base of tongue

C024 Lingual tonsil
Tonsils:
C090 Tonsillar fossa
C091 Tonsillar pillar
C098 Overlapping lesion of tonsil
C099 Tonsil NOS
C111 Adenoids/pharyngeal tonsil (does not include posterior wall of nasopharynx)

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.

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

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

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenoid cystic carcinoma 8200		
Polymorphous adenocarcinoma 8525	Cribriform adenocarcinoma Polymorphous low-grade adenocarcinoma Terminal duct carcinoma	
Squamous cell carcinoma 8070 ***Note 1***: Beginning 1/1/2022, keratinizing squamous cell carcinoma, HPV negative is coded 8086 for sites listed in Table 5 ***only***. A diagnosis of keratinizing squamous cell carcinoma, NOS is coded 8071. ***Note 2***: Beginning 1/1/2022, non-keratinizing squamous cell carcinoma, HPV positive is coded 8085 for sites listed in Table 5 ***only***. A diagnosis of non-keratinizing squamous cell carcinoma, NOS is coded 8072.	Conventional Squamous cell carcinoma NOS	Basaloid squamous cell carcinoma **8083** Keratinizing squamous cell carcinoma **8071 (see note 1)** Lymphoepithelial carcinoma **8082** Non-keratinizing squamous cell carcinoma **8072 (see note 2)** Papillary squamous cell carcinoma **8052** Squamous cell carcinoma HPV-negative **8086*** **Cases diagnosed prior to 1/1/2022:** ***Note:*** HPV-negative is not equivalent to HPV-mediated (p16-). According to the 2018 SEER Manual, HPV-type 16 refers to virus type and is different from p16 overexpression (p16+). HPV status is determined by tests designed to detect viral DNA or RNA. Tests based on ISH, PCR, RT-PCR technologies detect the viral DNA or RNA; whereas, the test for p16 expression, a surrogate marker for HPV, is IHC. HPV testing must be negative by viral detection tests in order to code histology as 8086. **Cases diagnosed 1/1/2022 forward:** ***Note:*** HPV mediated (p16-) test results can be used to assign code 8086. Squamous cell carcinoma HPV-positive **8085***

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
		Cases diagnosed prior to 1/1/2022: ***Note:*** HPV-positive is not equivalent to HPV-mediated (p16+). According to the 2018 SEER Manual, HPV-type 16 refers to virus type and is different from p16 overexpression (p16+). HPV status is determined by tests designed to detect viral DNA or RNA. Tests based on ISH, PCR, RT-PCR technologies detect the viral DNA or RNA; whereas, the test for p16 expression, a surrogate marker for HPV, is IHC. HPV testing must be positive by viral detection tests in order to code histology as 8085. **Cases diagnosed 1/1/2022 forward:** ***Note:*** HPV mediated (p16+) test results can be used to assign code 8085. Squamous cell carcinoma, spindle cell **8074** Verrucous carcinoma/Carcinoma cuniculatum **8051**

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

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 6: Tumors of Salivary Glands

Table 6 lists the more common histologies for the following head and neck subsites:

- **C079** Parotid gland, parotid NOS Stensen duct, parotid gland duct
- **C080** Submandibular gland, submaxillary gland, Wharton duct, submaxillary gland duct
- **C081** Sublingual gland; sublingual gland duct
- **C088** Overlapping lesion of major salivary glands
- **C089** Major salivary gland NOS; salivary gland NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.
Note: Hematopoietic neoplasms are common in the major salivary glands.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants**

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

Note 1: **Salivary duct carcinoma** was **assigned** code **8500** because it resembles high-grade duct carcinoma as found in the breast. These tumors are very aggressive. Code **8500 only** when the diagnosis is **exactly salivary duct carcinoma.**
Note 2: Assign code **8140** when the diagnosis is **salivary gland adenocarcinoma**.

Table begins on next page

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Acinic cell carcinoma 8550	ACC Acinar cell carcinoma Acinic cell adenocarcinoma	
Adenocarcinoma 8140	Adenocarcinoma NOS Unclassified adenocarcinoma Salivary gland adenocarcinoma NOS	Basal cell adenocarcinoma **8147** Basal cell adenocarcinoma-ex-monomorphic adenoma **8147** Malignant dermal analogue tumor **8147** Carcinoma ex-pleomorphic adenoma **8941** Clear cell carcinoma (CCC)/hyalinizing clear cell carcinoma **8310** Cribriform adenocarcinoma **8201** Intestinal-type adenocarcinoma **8144** Large cell carcinoma NOS/large cell undifferentiated carcinoma **8012** Lobular carcinoma **8520** Mucinous cystadenocarcinoma **8470** Mucoepidermoid carcinoma (MEC)/malignant mucoepidermoid tumor **8430** Papillary cystadenocarcinoma **8450** Polymorphous adenocarcinoma (PAC) **8525** Polymorphous low-grade adenocarcinoma **8525** Terminal duct carcinoma **8525** Salivary duct carcinoma **8500** Cribriform cystadenocarcinoma low-grade **8500/2** Ductal carcinoma/adenocarcinoma **8500** High-grade ductal carcinoma **8500** Intraductal carcinoma **8500/2** Intraductal carcinoma low-grade **8500/2** Undifferentiated carcinoma **8020**

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenoid cystic carcinoma 8200	ACC	
Carcinosarcoma 8980	Carcinosarcoma NOS True malignant mixed tumor	
Cystadenocarcinoma 8440		
Epithelial-myoepithelial carcinoma 8562	Adenomyoepithelioma	
Lymphoepithelial carcinoma (LEC) 8082	Lymphoepithelioma-like carcinoma Malignant lymphoepithelial lesion Undifferentiated carcinoma with lymphoid stroma	
Myoepithelial carcinoma 8982	Malignant myoepithelioma	
Neuroendocrine carcinoma 8246	Neuroendocrine carcinoma NOS	Large-cell neuroendocrine carcinoma **8013** Small cell carcinoma NOS/small cell neuroendocrine carcinoma **8041**
Oncocytic carcinoma 8290	Malignant oncocytoma Oncocytic adenocarcinoma	
Sebaceous adenocarcinoma 8410	Sebaceous carcinoma. NOS	
Secretory carcinoma 8502*	Mammary analog secretory carcinoma	
Squamous cell carcinoma 8070	Conventional Squamous cell carcinoma NOS SCC Squamous carcinoma Squamous cell carcinoma NOS	

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

Table 7: Tumors of Odontogenic and Maxillofacial Bone (Mandible, Maxilla)

Table 7 lists the more common histologies for the following head and neck subsites:

C410 Bones of skull and face and associated joints; maxilla
C411 Mandible; jaw bone NOS; lower jaw bone; temporomandibular joint

Note: The term odontogenic means originating in tooth forming tissue and bone. Code the primary site listed on the pathology report. The common primary sites include the maxillofacial skeleton (**C410** maxilla and **C411** mandible)

There are no hematopoietic neoplasms common to odontogenic bone or tissue. If a hematopoietic neoplasm such as lymphomas, myelomas, plasmacytoma etc., is diagnosed, verify the primary site. If the primary site is correct, see the Hematopoietic Database.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants**

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

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including osteosarcoma 9180/3 (column 3). Osteosarcoma is also a NOS because it has subtypes/variants. The subtypes/variants are indented under the NOS (osteosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that osteosarcoma has subtypes/variants.

When using the Solid Tumor Rules, osteosarcoma and its subtypes/variants are treated the same as all NOS and subtypes/variants.

Table begins on next page

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Ameloblastic carcinoma-primary type 9270/3	AC Ameloblastic carcinoma Ameloblastic carcinoma, dedifferentiated Ameloblastic carcinoma, secondary type Primary intraosseous carcinoma NOS (PIOC) Sclerosing odontogenic carcinoma (SOC)	Metastasizing ameloblastoma **9310/3** ***Note:*** This is an ameloblastoma which has a benign appearance but metastasizes
Clear cell odontogenic carcinoma 9341* ***Note:*** **Clear cell** odontogenic tumors **were** classified as **benign prior** to the **2005** edition of **WHO** ***Pathology & Genetics Head and Neck Tumors***	CCOC	
Ghost cell odontogenic carcinoma 9302*	Aggressive epithelial ghost cell odontogenic tumor Calcifying ghost cell odontogenic carcinoma Carcinoma arising in calcifying odontogenic cyst Malignant calcifying ghost cell odontogenic tumor Malignant calcifying odontogenic cyst Malignant epithelial odontogenic ghost cell tumor	

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Odontogenic carcinosarcoma 8980/3	Ameloblastic carcinosarcoma Malignant odontogenic mixed tumor Mixed odontogenic carcinoma	Odontogenic sarcoma/ameloblastic fibrosarcoma **9330/3**
Sarcoma NOS 8800/3 ***Note 1:*** Osteosarcoma **9180/3** has subtypes/variants: Chondroblastic osteosarcoma **9181/3** Intraosseous well-differentiated osteosarcoma/low-grade central osteosarcoma **9187/3** Parosteal osteosarcoma **9192/3** Periosteal osteosarcoma **9193/3** ***Note 2:*** Chondrosarcoma grade 2/3 **9920/3** has a subtype/ variant: Mesenchymal chondrosarcoma **9240/3**		Chondrosarcoma grade 2/3 **9220/3** Mesenchymal chondrosarcoma **9240/3** Osteosarcoma/osteogenic sarcoma **9180/3** Chondroblastic osteosarcoma **9181/3** Intraosseous well-differentiated osteosarcoma/low-grade central osteosarcoma **9187/3** Parosteal osteosarcoma **9192/3** Periosteal osteosarcoma **9193/3**

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

Table 8: Tumors of Ear

Table 8 lists the more common histologies for the following head and neck subsites:

- **C301** Middle ear; inner ear; auditory tube; eustachian tube; mastoid antrum; tympanic cavity

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the Hematopoietic Database.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

Specific or NOS Term and Code	Synonyms
Endolymphatic sac tumor 8140 ***Note:*** The endolymphatic sac is located within the inner ear C301.	Adenocarcinoma Heftner tumor Low-grade papillary adenocarcinoma of endolymphatic sac origin
Squamous cell carcinoma of the middle ear 8070 ***Note:*** This neoplasm **arises** in the squamous epithelium within the middle ear C301.	SCC Squamous carcinoma Squamous cell carcinoma NOS

Table 9: Paraganglioma of Carotid Body, Extra-adrenal, Larynx, Middle Ear, Vagal Nerve

Table 9 lists codes for paragangliomas diagnosed prior to 1/1/2021 and new codes for cases diagnosed 1/1/2021 forward. Table 9 does not list all paragangliomas, only those common to head and neck sites.

Cases diagnosed prior to 1/1/2021:
Only report these neoplasms when the pathology/tissue specifies malignant (/3) behavior. Change the behavior using ICD-O-3 Rule F Matrix Concept.

Cases diagnosed 1/1/2021 forward:
The term "malignant" is no longer required to assign malignant (/3) behavior. Paragangliomas diagnosed 1/1/2021 or after are malignant unless otherwise stated by the pathologist.

Coding Primary Site:
Paragangliomas have a separate chapter in the WHO Classification of Head and Neck Tumors which is why they are included in the Head and Neck Solid Tumor Rules. Some variants of paraganglioma are specific to certain sites but may occur in sites other than the nervous system. Variants that have specific sites are noted with the appropriate C-code in Table 9. Always code the site noted by the physician. If site is not stated or unclear and histology term does not have a specific site noted in Table 9, code to autonomic nervous system **C479**.

Definitions

- **Ganglion:** A group of nerve cell bodies located outside the central nervous system.
- **Sympathetic nervous system:** It is a part of the autonomic nervous system and contains adrenergic fibers which depress secretion, decrease tone and contractility of smooth muscle and increase heart rate.

Column 1 lists **ICD-O histology** term or NOS term and C-code if appropriate
Column 2 lists **ICD-O code** for cases **diagnosed prior to 1/1/2021** and stated to be malignant (/3)
Column 3 lists **ICD-O code** for cases **diagnosed 1/1/2021 forward**
Column 4 lists **synonyms** for the specific term. Synonyms have the **same ICD-O code** as the specific term.

Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Term and Code	ICD-O Code DX prior to 1/1/2021 *Must be stated to be malignant*	ICD-O Code DX 1/1/2021 forward *"Malignant" no longer required to assign /3*	Synonyms (Per ICD-O-3.2)
Aortic body paraganglioma (C75.5)	8691/3	8691/3	Aortic body tumor Aorticopulmonary paraganglioma
Carotid body paraganglioma (C75.4)	8692/3	8692/3	Carotid body tumor
Extra-Adrenal paraganglioma, NOS	8693/3	8693/3	Nonchromaffin paraganglioma, NOS Chemodectoma Composite paraganglioma
Laryngeal paraganglioma	8690/3	8693/3	
Middle ear paraganglioma (C75.5)	8690/3	8690/3	Glomus jugulare tumor Jugular Jugulotympanic paraganglioma
Paraganglioma, NOS	8680/3	8680/3	
Parasympathetic paraganglioma	8682/3	8682/3	
Sympathetic paraganglioma	8681/3	8681/3	
Vagal paraganglioma ***Note:*** Vagal paraganglioma has the same histology code as laryngeal paraganglioma. Extra-adrenal, laryngeal and vagal are in separate rows to emphasize primary site.	8690/3	8693/3	

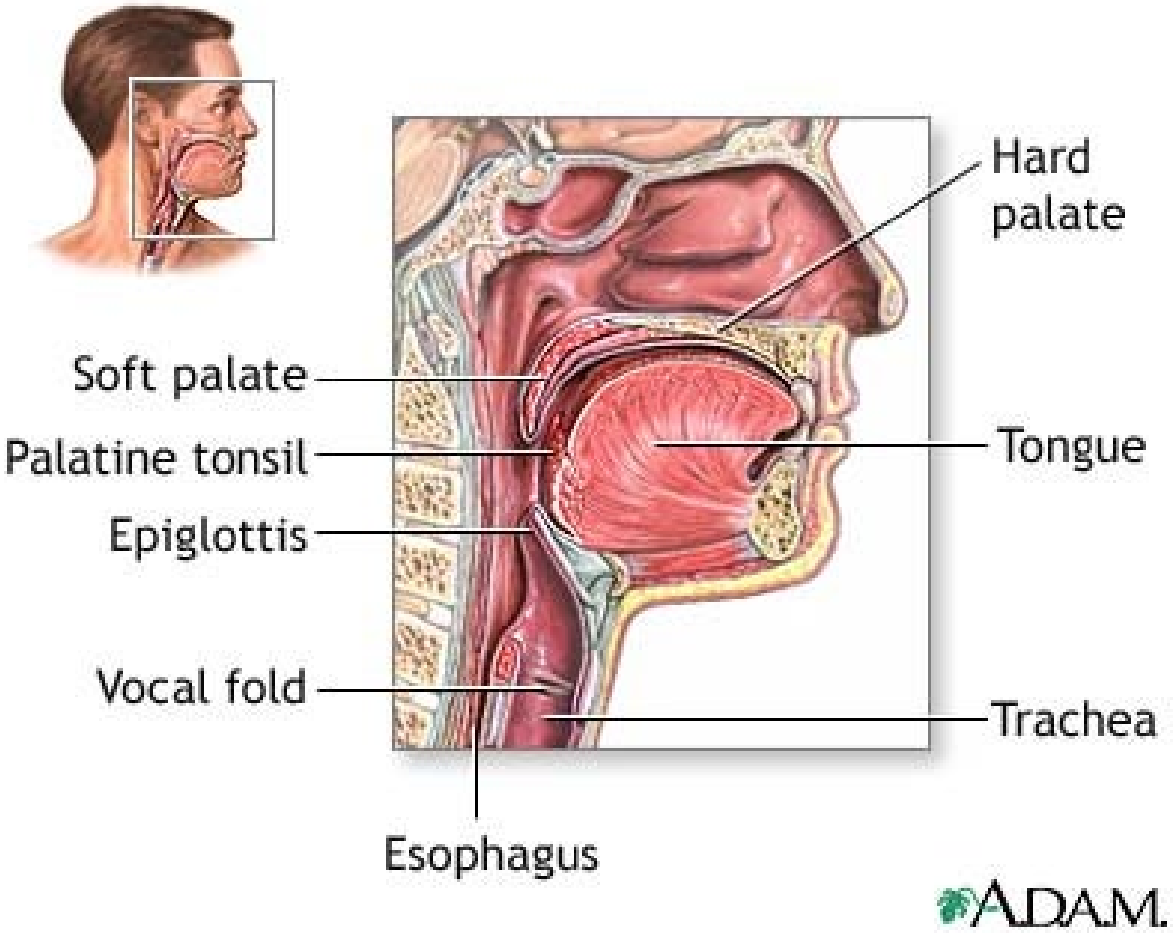
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Table 10: Paired Sites

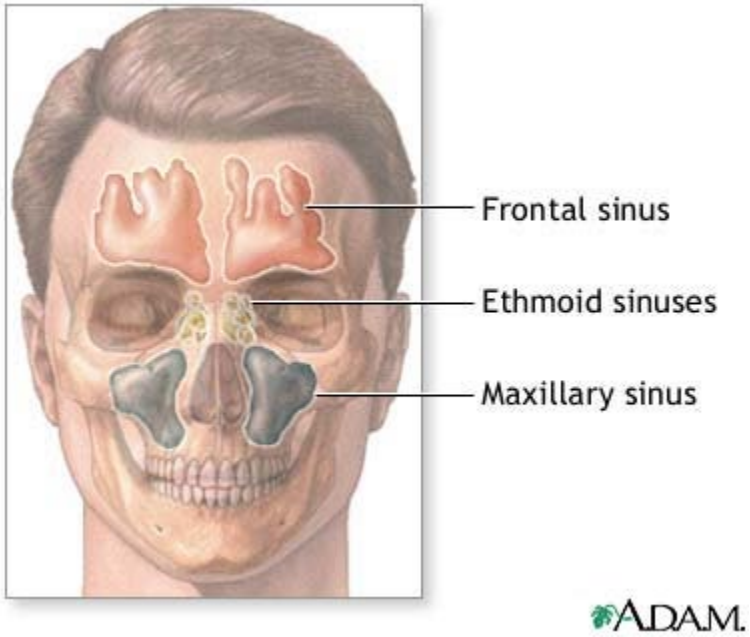
Laterality **must be coded** for all of the following sites. SEER does allow coding laterality for sites not listed in **Table 10.**

Paired Sites	Site Code
Carotid body	C754
Frontal sinus	C312
Maxillary sinus	C310
Middle ear	C301
Nasal cavity (excluding nasal cartilage, nasal septum)	C300
Tonsil	C098, C099
Parotid gland	C079
Sublingual gland	C081
Submandibular gland	C080

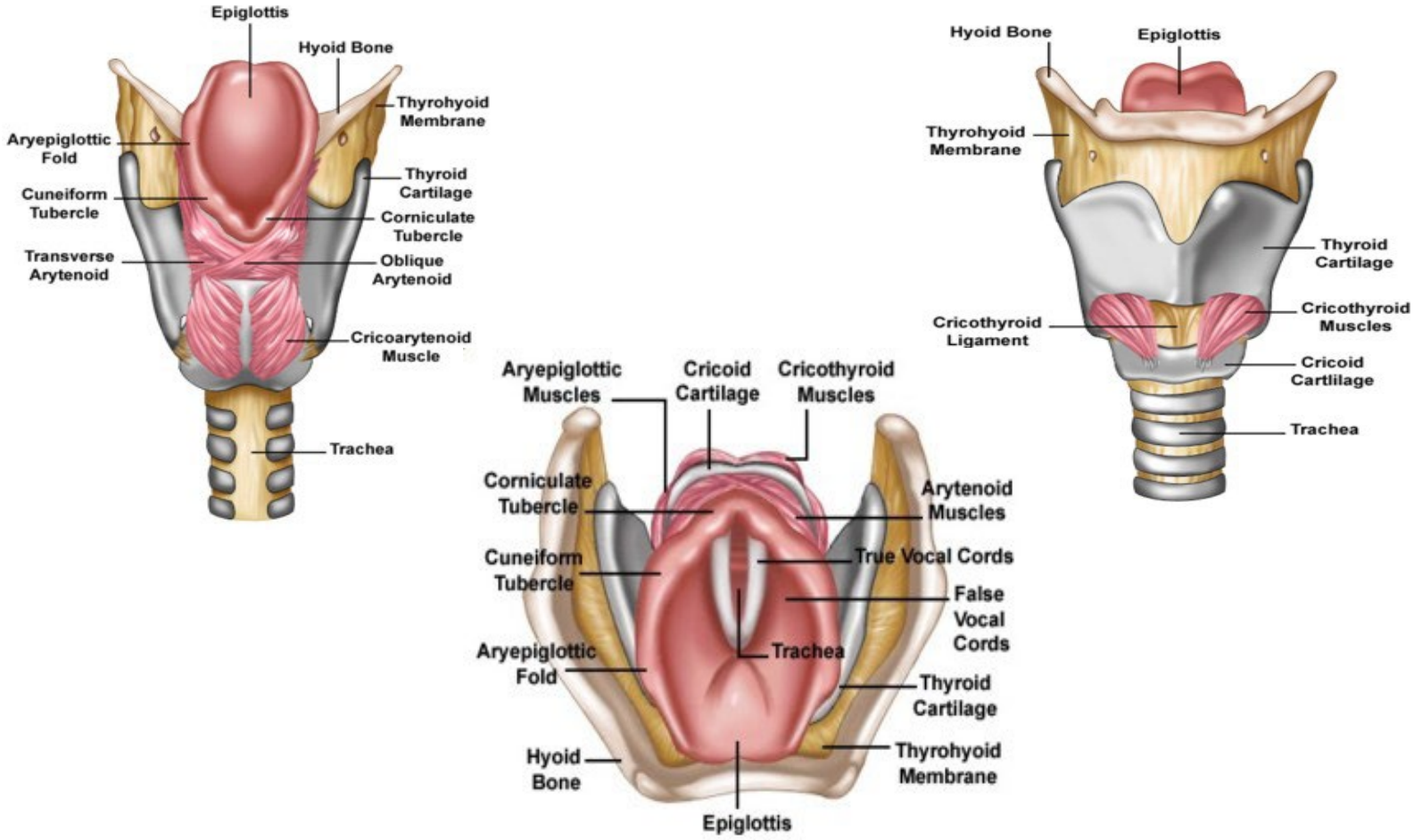
Illustrations



Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



Nasal Sinuses



Larynx

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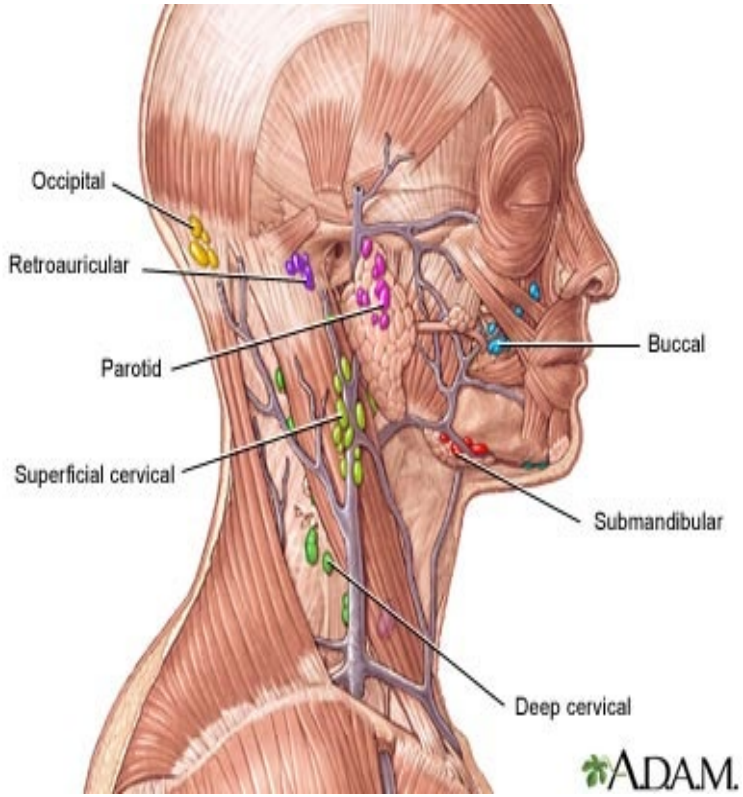
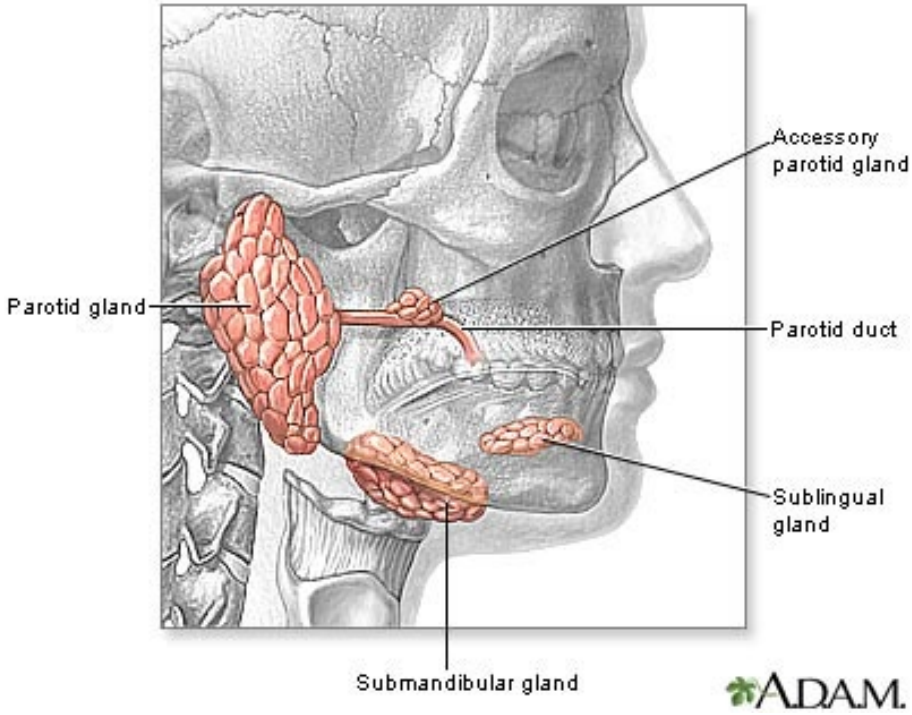
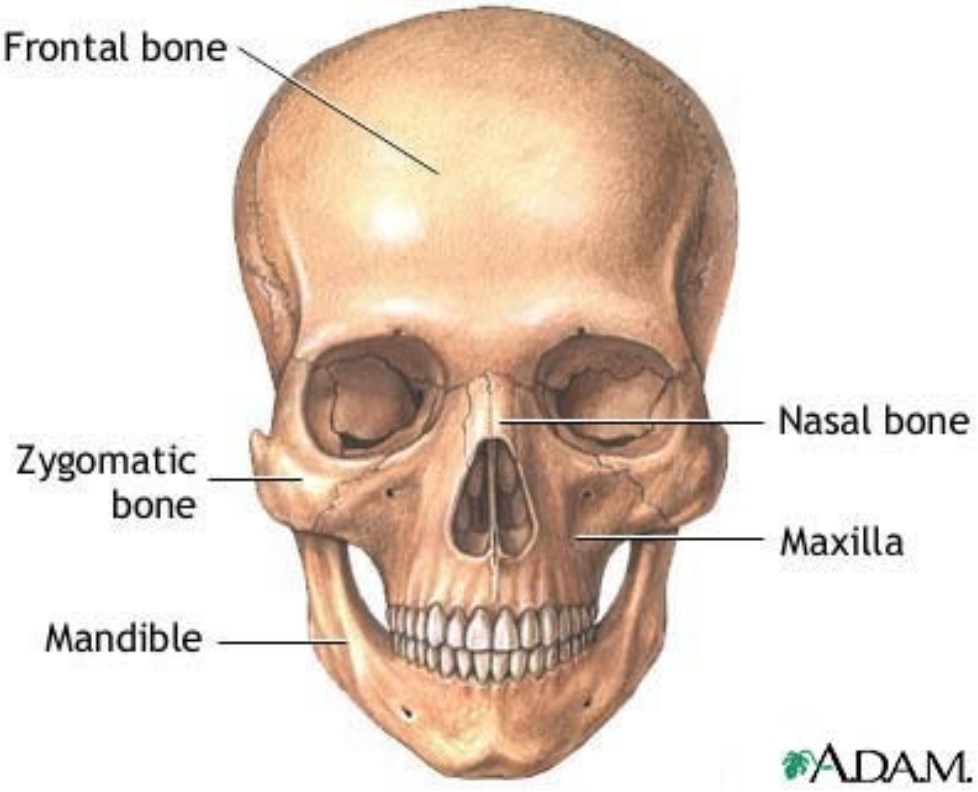
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



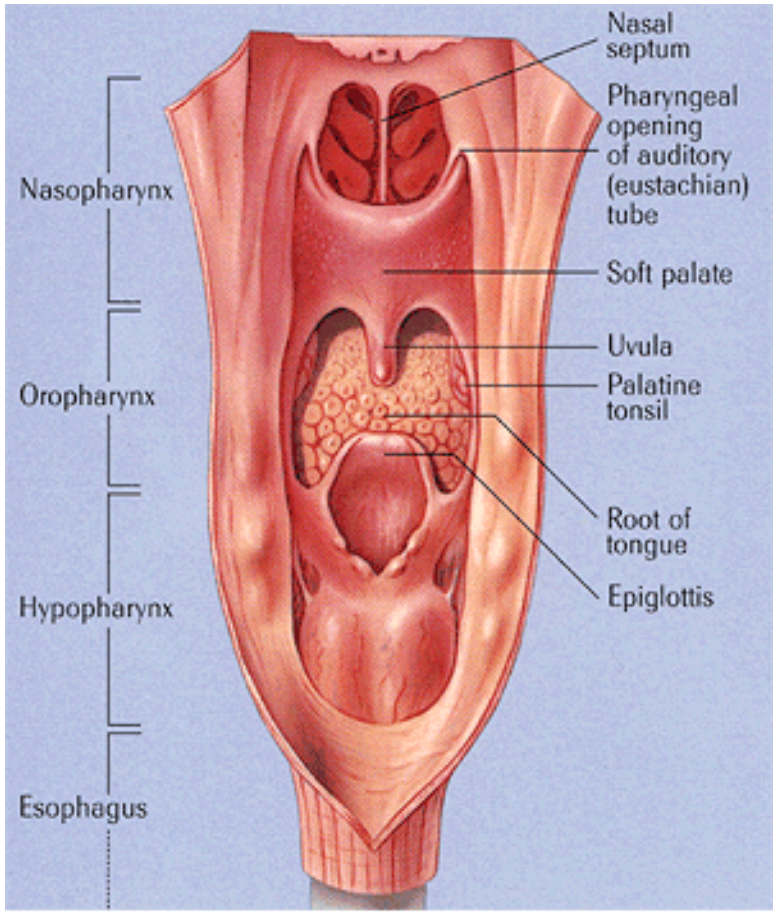
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



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Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Bone marrow
- Discontinuous lesions/nodules in soft tissue adjacent to primary site
- Regional and distant lymph nodes for the primary site being abstracted as identified in Summary Staging Manual
- Liver
- Lung
- Skin

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on date of diagnosis.

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

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor **or multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Pathology reports which do not specify whether a single tumor or multiple tumors were biopsied and/or resected

Example 1: History and physical exam states large tumor in nasopharynx. Biopsy base of tongue shows squamous cell carcinoma. No further information available. Abstract a single primary.

Example 2: Hospital A reports a biopsy of the upper lip mucosa. Hospital B reports a biopsy of the commissure of the lip. There is no information on whether this is a single tumor or whether there are separate tumors. Code a single primary.

This is the end of instructions for Unknown if Single or Multiple Tumors.

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

Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

IMPORTANT: If the current tumor was **preceded** by a tumor in the same primary site, go to the **Multiple Tumors module.**

Rule M2 Abstract a **single primary**[i] when there is a **single tumor**.
Note 1: A single tumor is <u>always</u> a single primary.
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor.

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

Multiple Tumors

Note: Multiple tumors may be a single primary or multiple primaries.

Rule M3 Abstract **multiple primaries**[ii] when there are separate/non-contiguous tumors in any two of the following sites:

- Glottis **C320 AND/OR** supraglottis **C321 AND/OR** subglottis **C322** A**ND/OR** laryngeal cartilage **C323**
- Hard palate **C050 AND/OR** soft palate **C051 AND/OR** uvula **C052**
- Maxilla **C410 AND** Mandible **C411**
- Maxillary sinus **C310 AND/OR** ethmoid sinus **C311 AND/OR** frontal sinus **C312 AND/OR** sphenoid sinus **C313**
- Nasal cavity **C300 AND** middle ear **C301**
- Postcricoid **C130 AND/OR** hypopharyngeal aspect of aryepiglottic fold **C131 AND/OR** posterior wall of hypopharynx **C132**
- Submandibular gland **C080 AND** sublingual gland **C081**
- Upper gum **C030 AND** lower gum **C031**
- Upper lip **C000** or **C003 AND** lower lip **C001** or **C004**

Note 1: Use this rule only for **multiple tumors.**
Note 2: Timing is irrelevant.
Note 3: Histology is irrelevant.
Note 4: These primary sites differ at the fourth character of the site code Cxx**X**. Use this rule **ONLY** for the primary sites listed.

Rule M4 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are present in sites with ICD-O **site** codes that **differ** at the second C**X**xx, and/or third characters Cx**X**x.
Note 1: Use this rule only for **multiple tumors.**
Note 2: Timing is irrelevant.
Note 3: Histology is irrelevant.

Rule M5 Abstract **multiple primaries**[ii] when there are separate/non-contiguous tumors on both the **right** side and the **left** side of a paired site.
Note 1: See **Table 10** for a list of **paired sites.**
Note 2: Use this rule only for **multiple tumors.**
Note 3: Timing is irrelevant.
Note 4: Histology is irrelevant.

Rule M6 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being **clinically disease-free** for **greater than five years** after the original diagnosis or last recurrence.
Note 1: **Clinically** disease-free means that there was **no evidence** of recurrence on follow-up.
- Scopes are NED
- Scans are NED

Note 2: When there is a recurrence less than or equal to five years of diagnosis, the **"clock"** starts over. The time interval is calculated from the **date of last recurrence.** In other words, the patient must have been disease-free for greater than five years from the date of the last recurrence.
Note 3: When it is **unknown/not documented** whether the patient had a recurrence, use **date of diagnosis** to compute the time interval.
Note 4: When the patient has more than one Head & Neck primary, it is often difficult to determine which primary recurred. Use the last date of recurrence for any tumor to calculate the time interval.
Note 5: The physician may state this is a **recurrence,** meaning the patient had a previous head and neck tumor and now has another head and neck tumor. **Follow the rules**; do not attempt to interpret the physician's statement.

Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M7 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3 of the appropriate site table (**Tables 1-9**) in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS**: Alveolar rhabdomyosarcoma 8920/3 and embryonal rhabdomyosarcoma 8910/3 are both subtypes of rhabdomyosarcoma 8900/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Colloid-type adenocarcinoma 8144 is a subtype of adenocarcinoma NOS 8140; Spindle cell squamous cell carcinoma 8074 is a subtype of squamous cell carcinoma 8070. They are distinctly different histologies. Abstract multiple primaries.

Rule M8 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are on **different rows** in the appropriate site table (**Tables 1-9**) in the Equivalent Terms and Definitions. Timing is irrelevant.

Note: Each row in the table is a **distinctly different** histology.

Rule M9 Abstract a **single primary**[i] (the invasive) when an **in situ** tumor is diagnosed **after** an **invasive** tumor **in the same primary site**.

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.

Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS. See **Tables 1-9** in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

Note 3: Do **not** change **date of diagnosis**.

Note 4: If the case has already been submitted to the central registry, **report** all changes.

Note 5: The physician **may stage both** tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

Note 6: See the **COC** and SEER manuals for instructions on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M10 Abstract a **single primary**[i] (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60** days **after** an **in situ** tumor **in the same primary site**.

Note 1: The rules are **hierarchical.** Only use this rule when none of the previous rules apply.

Note 2: The tumors may be an **NOS** and a **subtype/variant** of that NOS.

Note 3: When the case has been abstracted, **change behavior** code on original abstract from /2 to /3. Do **not** change **date of diagnosis**.

Note 4: If the case has already been submitted to the central registry, **report** all changes.
Note 5: The physician may **stage both** tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).
Note 6: See the **COC** and **SEER manuals** for instructions on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M11 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor.
Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: Abstract **both** the invasive and in situ tumors.
Note 3: Abstract as multiple primaries even if **physician states** the invasive tumor is disease **recurrence** or **progression.**
Note 4: This rule is based on long-term epidemiologic studies of recurrence intervals. The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were also authors, co-authors, or editors of the AJCC Staging Manual.

Rule M12 Abstract a **single primary**[i] when separate/non-contiguous tumors **in the same primary site** are on **the same row** in the appropriate site table **(Tables 1-9)** in the Equivalent Terms and Definitions. Timing is irrelevant.
Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3) **OR**
- A NOS histology in column 3 with an indented subtype/variant

Rule M13 Abstract a **single primary**[i] when none of the previous rules apply.
Note: Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

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[i] Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.
[ii] Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted

Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

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

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

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

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

Code the **most specific histology** from either **resection** or **biopsy**.

Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

This is a hierarchical list of source documentation.

1. **Tissue or pathology report from biopsy or resection of primary site** (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol

   Note 1: Addendums and comments on the pathology report are given a high priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
   Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.
   Note 3: The CAP protocol is a checklist which:

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

2. Cytology of primary site (fine needle aspirate (FNA))
3. Tissue/pathology from a **metastatic** site
   Note 1: Code the behavior /3
   Note 2: The tissue from a metastatic site often shows **variations** from the primary tumor. When it is the only tissue available, it is **more accurate** than a **scan**.
   Note 3: This includes cytology from a regional lymph node.
4. **Scan:** The following list is in **priority** order.
   A. **CT**
   B. **MRI**
   C. **PET**
5. Code the histology **documented** by the physician when none of the above are available. Use the **documentation** in the following **priority order:**
   A. Treatment plan
   B. Tumor Board
   C. Documentation in the medical record that **refers to original pathology, cytology, or scan(s)**
   D. Physician's **reference to** type of cancer (**histology**) in the medical record
   Note 1: Code the specific histology when documented.
   Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented.

Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Coding Histology

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

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

   Example 1: Diagnosis for a single tumor is adenocarcinoma 8140 with the majority or predominant part of tumor being enteric-type adenocarcinoma 8144. Code the subtype/variant: enteric-type adenocarcinoma 8144.
   Example 2: Diagnosis for a single tumor is squamous cell carcinoma 8070 with minority of tumor being spindle cell squamous cell carcinoma 8074. Code the subtype/variant: spindle cell squamous cell carcinoma 8074.
   Example 3: Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.

   Note 1: The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.
   Example: When the diagnosis is adenocarcinoma with an enteric-type **adenocarcinoma** component, code enteric-type adenocarcinoma 8144.
   Negative Example: When the diagnosis is simply adenocarcinoma with an enteric-type component, code adenocarcinoma NOS 8140. Do not assume this is enteric-type adenocarcinoma. This could be enteric-type differentiation or features.

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

2. Code the histology 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".
   ***Note*:** Do not code differentiation or features when there is no specific ICD-O code.

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

      Example: Outpatient biopsy says probably squamous cell carcinoma HPV-negative. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology squamous cell carcinoma HPV-negative. The case meets the criteria in #**3A.**

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

      Example 1: The pathology diagnosis is adenocarcinoma consistent with intestinal type adenocarcinoma. The oncology consult says the patient has intestinal type adenocarcinoma of the sinonasal tract. This is clinical confirmation of the diagnosis, code intestinal type adenocarcinoma. The case meets the criteria in **bullet 1**.

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

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

List of Ambiguous Terminology

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

4. **Do not code** histology when described as:

   - Architecture
   - Foci; focus; focal
   - Pattern

Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, C479, C754, C755
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule H1 Code the histology when only **one histology** is present.

Note 1: Use **Tables 1-9** to code histology. New codes, terms, and synonyms are included in **Tables 1-9** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Tables 1-9,** use the **ICD-O** and all **updates**.

Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Tables 1-9, ICD-O or all updates.

Note 4: **Cases diagnosed prior to 1/1/2022:** Squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086) are coded only when HPV status is determined by tests based on ISH, PCR, RT-PCR technologies to detect the viral DNA or RNA. p16 is not a valid test to assign these histology codes.
Cases diagnosed 1/1/2022 forward: p16 test results can be used to code squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086).

Rule H2 Code the **invasive** histology when **in situ** and **invasive** histologies are present in the **same tumor.**

***Example*:** The tissue/pathologic diagnosis is invasive squamous cell carcinoma **8070/3** and keratinizing squamous cell carcinoma in situ **8071/2**. Code the invasive histology, SCC **8070/3** even though it is not the most specific histology.

Rule H3 Code the subtype/variant when there is a **NOS** and a **single subtype/variant** of that NOS such as the following:

- Adenocarcinoma/endolymphatic sac tumor **8140** and a subtype/variant of adenocarcinoma
- Ameloblastic carcinoma primary type **9270** and a subtype variant of ameloblastic carcinoma primary type
- Chondrosarcoma grade 2/3 **9220** and a subtype/variant of chondrosarcoma grade 2/3
- Neuroendocrine carcinoma **8246** and a subtype/variant of neuroendocrine carcinoma
- Odontogenic carcinosarcoma **8980** and a subtype/variant of odontogenic carcinosarcoma
- Sarcoma **8800/3** and a subtype/variant of sarcoma
- Squamous cell carcinoma **8070** and subtype/variant of squamous carcinoma
- Well differentiated neuroendocrine carcinoma **8240** and a subtype/variant of well differentiated neuroendocrine carcinoma

Note: See **Tables 1-9** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

This is the end of instructions for Single Tumor

Code the histology according to the rule that fits the case

Multiple Tumors Abstracted as a Single Primary

Note: Before coding histology, the **Multiple Primary Rules must be applied.**

Rule H4 Code the **histology** when only **one** histologic type is identified for **all tumors.**

Note 1: Use **Tables 1-9** to code histology. New codes, terms, and synonyms are included in **Tables 1-9** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Tables 1-9,** use the **ICD-O** and all **updates**.

Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Tables 1-9, ICD-O or all updates.

Rule H5 Code the **invasive** histology when one of the following criteria are met:

- All **tumors** have both **invasive and in situ** elements **OR**
- **One** or more tumors are **invasive** and **one** or more tumors are **in situ**

Note 1: **Multiple Primary Rules must be applied** to be certain all tumors are a single primary.

Note 2: When the NOS is invasive and the subtype/variant is situ, code the NOS (invasive).

Rule H6 Code the **subtype/variant** when **all** tumors are a **NOS** and a **single subtype/variant** of that NOS such as the following:

- Adenocarcinoma/endolymphatic sac tumor **8140** and a subtype/variant of adenocarcinoma
- Ameloblastic carcinoma primary type **9270** and a subtype variant of ameloblastic carcinoma primary type
- Chondrosarcoma grade 2/3 **9220** and a subtype/variant of chondrosarcoma grade 2/3
- Neuroendocrine carcinoma **8246** and a subtype/variant of neuroendocrine carcinoma
- Odontogenic carcinosarcoma **8980** and a subtype/variant of odontogenic carcinosarcoma
- Sarcoma **8800/3** and a subtype/variant of sarcoma
- Squamous cell carcinoma **8070** and subtype/variant of squamous carcinoma
- Well differentiated neuroendocrine carcinoma **8240** and a subtype/variant of well differentiated neuroendocrine carcinoma

Note: See **Tables 1-9** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

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

Code the histology according to the rule that fits the case

Kidney Equivalent Terms and Definitions
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Introduction

Note 1: Tables and rules refer to ICD-O rather than ICD-O-3. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Note 3: **Renal cell carcinoma** (RCC) **8312** is a **group term** for glandular (adeno) carcinoma of the kidney. Approximately 85% of all malignancies of the kidney C649 are RCC or subtypes/variants of RCC.

- See **Table 1** for renal cell carcinoma subtypes/variants.
- Clear cell renal cell carcinoma (ccRCC) 8310 is the most common subtype/variant of RCC.

Note 4: **Transitional cell carcinoma** rarely arises in the kidney C649. Transitional cell carcinoma of the upper urinary system usually arises in the renal pelvis C659. Only code a transitional cell carcinoma for kidney in the **rare** instance when **pathology confirms** the tumor originated in the kidney.

Note 5: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Biomarkers may identify the histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

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Changes from 2007 Rules

These changes are effective with cases diagnosed 1/1/2018 and later. WHO Classification of Tumors of the Urinary System and Male Genital Organs was published in 2016.

1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, 2018 Rules instruct "Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor)."
2. **New histology terms and codes were included** (identified by asterisks (*) in the histology table in the Terms and Definitions).
   A. Histologies with terms that indicate they are **hereditary** (hereditary leiomyomatosis and renal cell carcinoma syndrome–associated RCC **8311**)
   B. Histologies with **genetic anomalies** (succinate dehydrogenase–deficient RCC)
3. Some histologies are rare and are not listed in the tables; refer to ICD-O and all updates.
   Note: Renal cell spindle cell carcinoma 8318 is no longer a recommended term.

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Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
  Note: "And" and "with" are used as synonyms when describing multiple histologies within a single tumor.
- Multifocal; multicentric
- Carcinoma; adenocarcinoma
  - A histology type must be stated for these terms to be equal
  - Example: Renal cell carcinoma and renal cell adenocarcinoma are both coded 8312
- Simultaneous; existing at the same time; concurrent; prior to first course treatment
- Site; topography
- Tumor; mass; tumor mass; lesion, neoplasm
  - The terms tumor, mass, tumor mass, lesion, neoplasm and nodule are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms **unless** there is a **physician's statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to **determine** multiple **primaries**
  - **Do not** use these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

Terms that are NOT Equivalent or Equal

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

- Carcinoma, NOS 8010 is not equivalent to adenocarcinoma, NOS 8140
- **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**

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Table 1: Specific Histologies, NOS, and Subtypes/Variants

Use Table 1 as directed by the **Histology Rules** to assign the more common histology codes for kidney tumors.

Column 1 contains specific and NOS ICD-O histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants**

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

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including rhabdomyosarcoma 8900/3 (column 3). Rhabdomyosarcoma is also a NOS because it has subtypes/variants. The subtypes/variants are indented under the NOS (rhabdomyosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that rhabdomyosarcoma has subtypes/variants.

When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, rhabdomyosarcoma **8900/3** and alveolar type rhabdomyosarcoma **8920/3** are a NOS and a subtype/variant, **NOT** two different subtypes.

Table begins on next page.

Kidney Equivalent Terms and Definitions
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NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants
Medullary carcinoma **8510** ***Note***: Renal medullary carcinoma is a new term (previously called renal spindle cell carcinoma).	Medullary adenocarcinoma Renal medullary carcinoma SMARCB1-deficient medullary-like RCC SMARCB1-deficient undifferentiated RCC, NOS SMARCB1-deficent dedifferentiated RCC of other specific subtypes	
Nephroblastoma **8960**	Wilms tumor	
Neuroendocrine tumor (NET) **8240/3** ***Note***: Extra-adrenal paraganglioma, NOS reportable for cases diagnosed 1/1/2024 forward	Carcinoid [OBS] Well-differentiated neuroendocrine tumor	Extra-adrenal paraganglioma **8693/3*** Large cell neuroendocrine carcinoma/tumor **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Renal cell carcinoma NOS **8312** ***Note 1:*** WHO, IARC, and CAP agree that sarcomatoid carcinoma is a pattern of differentiation, not a specific subtype, of renal cell carcinoma. ***Note 2:*** Sarcomatoid is listed in the CAP Kidney protocol under the header "features."	Eosinophilic renal cell carcinoma Oncocytic renal cell carcinoma RCC Sarcomatoid carcinoma Sarcomatoid renal cell carcinoma Succinate dehydrogenase-	Acquired cystic disease-associated renal cell carcinoma/tubulocystic renal cell carcinoma **8316*** Chromophobe renal cell carcinoma (ChRCC)/Hybrid oncocytic chromophobe tumor **8317** Clear cell papillary renal cell carcinoma **8323/3** ***Note:*** The 2016 WHO 4th Edition Classification of Tumors of the Urinary System and Male Genital Organs has reclassified this histology as a /1 because it is low nuclear grade and is now thought to be a neoplasia. This change has **NOT** yet been implemented and it **remains reportable.**

Kidney Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants
Note 3: Continue coding sarcomatoid renal cell carcinoma as 8312 until otherwise indicated. ***Note 4:*** "Oncocytic" indicates cells that have abundant eosinophilic cytoplasm due to the accumulation of mitochondria and is not a histologic type unless listed in column 3. ***Note 5:*** Beginning with cases diagnosed 1/1/2022 forward, SDHD is coded 8311/3. Cases diagnosed prior to 1/1/2022 should be coded 8312.	deficient renal cell carcinoma (SDHD) (pre-2022) Unclassified renal cell carcinoma	Clear cell renal cell carcinoma (ccRCC) **8310** Collecting duct carcinoma **8319** ELOC (formerly TCEB1) mutated RCC **8311^** Eosinophilic solid and cystic RCC **8311^** Fumarate hydratase-deficient RCC ALK-rearranged RCC **8311^** Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma **8311^** MiT family translocation renal cell carcinomas **8311^** Succinate dehydrogenase-deficient renal cell carcinoma (SDHD) **8311^ (reportable beginning 1/1/2022)** T(6;11) RCC **8311^** TFE3-rearranged RCC **8311^** TFEB-altered RCC **8311^** Xp11 translocation RCC **8311^** ***Note*:** The 8311 terms marked with a caret^ above have the same ICD-O code but are distinctly different histologies. Because they are different, they are on different lines in column 3 (see M rules). Mucinous tubular and spindle cell carcinoma **8480*** Papillary renal cell carcinoma (PRCC) **8260**

Kidney Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants
Sarcoma **8800/3** ***Note*: Rhabdomyosarcoma** is a NOS with the following subtype/variants: Alveolar rhabdomyosarcoma **8920** Embryonal rhabdomyosarcoma **8910** Pleomorphic rhabdomyosarcoma **8901** Spindle cell/sclerosing rhabdomyosarcoma **8912**		Angiosarcoma **9120/3** Clear cell sarcoma/bone-metastasizing renal tumor of childhood **8964/3** Leiomyosarcoma/renal vein leiomyosarcoma **8890/3** Osteosarcoma **9180/3** Primitive/peripheral neuroectodermal tumor (pNET)/Ewing sarcoma **9364/3** Rhabdomyosarcoma **8900/3** Alveolar rhabdomyosarcoma **8920/3** Embryonal rhabdomyosarcoma **8910/3** Pleomorphic rhabdomyosarcoma **8901/3** Spindle cell/sclerosing rhabdomyosarcoma **8912/3** Synovial sarcoma **9040/3**

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

Kidney Equivalent Terms and Definitions
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Table 2: Neoplasms which are Not Reportable

Column 1 lists the not reportable histology **term** and **code**. Not all of the non-reportable neoplasms have codes.
Column 2 lists **synonyms** for the term in column 1. Synonyms have the same histology code (if applicable) as listed in column 1.

Not Reportable Histology Term and Code	Synonyms
Adult cystic teratoma **8959/0**	Mixed epithelial and stromal tumor Pediatric cystic nephroma Renal epithelial stromal tumor
Angiomyolipoma **8860/0**	Angiomyolipoma with epithelial cysts Oncocytic angiomyolipoma
Congenital mesoblastic nephroma **8960/1**	CMN Cellular congenital mesoblastic nephroma Classic congenital mesoblastic nephroma Mesoblastic nephroma Mixed congenital mesoblastic nephroma
Cystic partially-differentiated nephroblastoma **8959/1**	
Epithelioid angiolipoma **8860/1***	
Hemangioblastoma **9161/1**	
Hemangioma **9120/0**	
Juxtaglomerular cell tumor **8361/0**	Functioning juxtaglomerular cell tumor Non-functioning juxtaglomerular cell tumor
Leiomyoma **8890/0**	
Lymphangioma **9170/0**	
Metanephric adenofibroma **9013/0**	Nephrogenic adenofibroma
Metanephric adenoma **8325/0**	
Metanephric stromal tumor **8935/1**	
Multilocular cystic renal neoplasm of low malignant potential **8316/1***	
Nephrogenic rests **(no code)**	

Kidney Equivalent Terms and Definitions
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

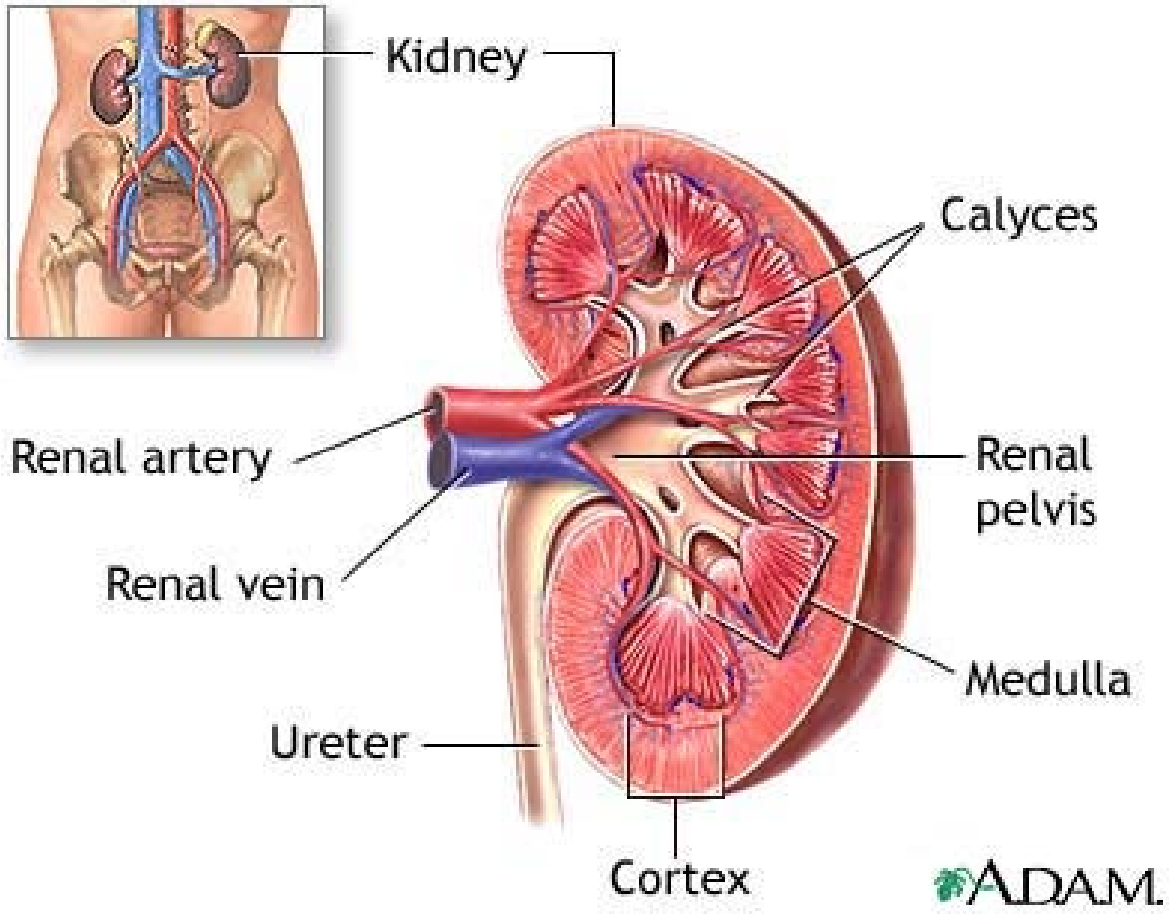
Not Reportable Histology Term and Code	Synonyms
Oncocytic tumor, NOS **(no code)**	
Oncocytoma **8290/0**	
Ossifying renal tumor of infancy **8967/0**	
Papillary adenoma **8260/0**	Tubulopapillary adenoma
Paraganglioma **8700/0** (non-reportable through 12/31/2023. See Table 1).	Extra-adrenal pheochromocytoma
Pediatric cystic nephroma **8959/0**	
Renomedullary interstitial cell tumor **8966/0**	Medullary fibroma
Schwannoma **9560/0**	
Solitary fibrous tumor **8815/1**	

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

Kidney Equivalent Terms and Definitions
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Illustrations

Kidney Anatomy (Includes Renal Pelvis)



Kidney Equivalent Terms and Definitions
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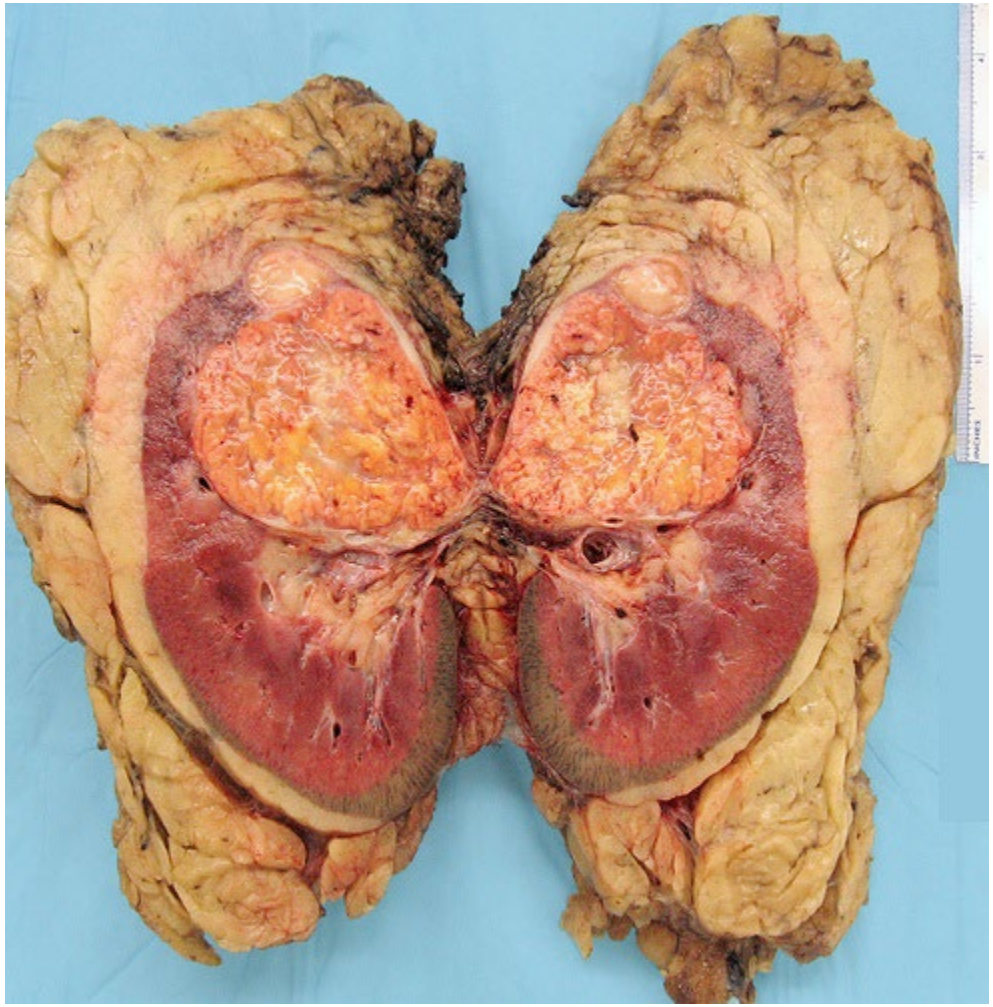
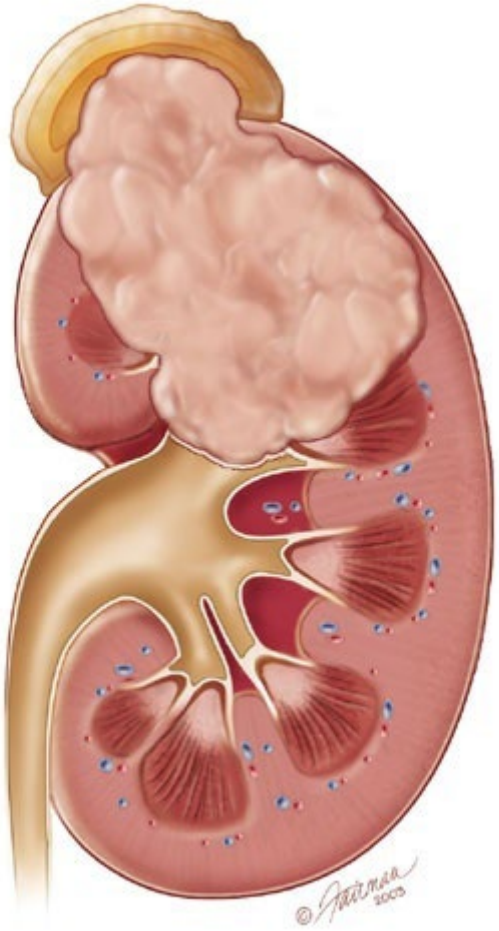
Pathology Specimen Kidneys



Kidney Equivalent Terms and Definitions
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Kidney Cancer



Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Adrenal gland
- Bones
- Bowel
- Brain
- Discontinuous nodules in surrounding tissue
- Regional and distant lymph nodes as identified in Summary Staging Manual
- Liver
- Lung

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown If Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor **or multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors.

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

Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule M2 Abstract a **single primary**[i] when there is a **single tumor**.
Note 1: A single tumor is always a single primary.
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor.

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

Multiple Tumors

Note: Multiple tumors may be a **single** primary **OR multiple** primaries.

Rule M3 Abstract **multiple primaries**[ii] when **multiple tumors** are present in sites with ICD-O **site** codes that **differ** at the second (C**X**xx), third (Cx**X**x) and/or fourth characters (Cxx**X**).
Note: When codes differ at the second, third, or fourth characters, the tumors are in different primary sites.

Rule M4 Abstract a **single primary**[i] when there are **bilateral nephroblastomas** (previously called Wilms tumors).
Note: Timing is irrelevant**;** the tumors may occur simultaneously OR the contralateral tumor may be diagnosed later (no time limit).

Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M5 Abstract **multiple primaries**[ii] when there are tumors in **both** the **right kidney** and in the **left kidney**. There may be:

- A single tumor in each kidney
- A single tumor in one kidney and multiple tumors in the contralateral kidney
- Multiple tumors in both kidneys

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.

Note 2: **ONLY** abstract a single primary when **pathology** proves the tumor(s) in one kidney is/are **metastatic** from the other kidney.

Rule M6 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being clinically disease-free for greater **than three years** after the original diagnosis or last recurrence.

Note 1: Clinically disease-free means that there was **no <u>evidence</u>** of recurrence on follow-up.

- Scans are NED
- Urine cytology is negative
- All other work-up is NED

Note 2: When there is a **recurrence** less than or equal to three years of diagnosis, the **"clock" starts over**. The time interval is calculated from the date of last recurrence. In other words, the patient must have been disease-free for greater than three years from the date of the last recurrence.

Note 3: When it is unknown/not documented whether the patient had a recurrence, **default** to **date of diagnosis** to compute the time interval.

Note 4: The physician may state this is a **recurrence,** meaning the patient had a previous kidney tumor and now has another kidney tumor. **Follow the rules;** do not attempt to interpret the physician's statement.

Note 5: The location and histology of the subsequent tumor is irrelevant. Kidney tumors that occur more than 3 years apart are always multiple primaries.

Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M7 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3, Table 1 in the Equivalent Terms and Definitions.

Note 1: The tumors may be subtypes/variants of the **same** or **different** NOS histologies.

- **Same NOS**: Clear cell renal cell carcinoma (ccRCC) 8310/3 and papillary renal cell carcinoma 8260/3 are both subtypes of renal cell carcinoma NOS 8312/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Pleomorphic rhabdomyosarcoma 8901/3 is a subtype/variant of rhabdomyosarcoma 8900/3; large cell neuroendocrine carcinoma 8013/3 is a subtype of small cell neuroendocrine tumor 8041/3. They are distinctly different histologies. Abstract multiple primaries.

Note 2: Abstract multiple primaries when you have any of the following combinations (all coded 8311):

- ELOC (formerly TCEB1) mutated RCC
- Eosinophilic solid and cystic RCC
- Fumarate hydratase-deficient RCC ALK-rearranged RCC
- Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma (HLRCC)
- MiT family translocation renal cell carcinoma
- Succinate dehydrogenase-deficient renal cell carcinoma (SDHD)
- t(6;11) RCC
- TFE3-rearranged RCC
- TFEB-altered RCC
- Xp11 translocation RCC

Rule M8 Abstract a **single primary**[i] when synchronous, separate/non-contiguous tumors are on the **same row** in Table 1 in the Equivalent Terms and Definitions. Tumors must be in the same kidney.

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code; see exception for 8311) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3) **OR**
- A NOS histology in column 3 with an indented subtype/variant

Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M9 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are on **different rows** in **Table 1** in the Equivalent Terms and Definitions.

Note: Each row in the table is a **distinctly different** histology.

Rule M10 Abstract a **single primary**[i] when an **in situ** tumor is diagnosed **after** an **invasive** tumor **AND** tumors occur in the same kidney.

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.

Note 2: The tumors **may** be a **NOS** and a **subtype/variant** of that NOS. See **Table 1** in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

Note 3: Once the patient has an invasive tumor, the **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.

Rule M11 Abstract a **single primary**[i] (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60** days **after** an **in situ** tumor in the same kidney.

Note 1: The rules are **hierarchical.** Only use this rule when none of the previous rules apply.

Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS.

Note 3: When the case has been abstracted, **change behavior** code on original abstract from /2 to /3.

Note 4: Do **not** change **date of diagnosis**.

Note 5: If the case has already been submitted to the central registry, **report** all changes.

Note 6: The physician may **stage both** tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

Note 7: See the **COC** and **SEER** manuals for **instructions** on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M12 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor.

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.

Note 2: Abstract **both** the invasive and in situ tumors.

Note 3: Abstract as multiple primaries even if **physician states** the invasive tumor is disease **recurrence** or **progression.**

Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M13 Abstract a **single primary**[i] when there are multiple tumors that **do not meet any** of the **above criteria.**

Note: Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

Example 1: Patient presents in 2018 with renal cell carcinoma in the right kidney. Patient has a history of a previous renal cell carcinoma in the right kidney diagnosed in 2016. This is a single primary because it is the same primary site and the same histology.

Example 2: Patient presents in 2020 with a clear cell renal cell carcinoma 8310/3 in the left kidney. The patient was diagnosed with renal cell carcinoma 8312/3 in 2018. This is a single primary because it is the same primary site and a NOS and subtype/variant of that NOS.

This is the end of instructions for Multiple Tumors.

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[i] Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

Kidney Histology Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

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

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

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

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

This is a hierarchical list of source documentation.

Code the **most specific** pathology/tissue from either **resection** or **biopsy**.
Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

1. Ti**ssue** or **pathology report from primary site** (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol
      Note 1: Addendums and comments on the pathology report are given the highest priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
      Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.

Note 3: The CAP protocol is a checklist which:

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

2. **Cytology** (urine)
3. Tissue/pathology from a **metastatic** site
   Note 1: Code the behavior /3.
   Note 2: The tissue from a metastatic **s**ite often shows **variations** from the primary tumor. When it is the only tissue available, it is more accurate than a scan.
4. **Scan:** The following list is **not in priority** order because they are not a reliable method for identifying specific **histology**(ies).
   A. MRI
   B. CT
   C. PET
5. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following priority order:
   A. Treatment plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that refers to original pathology, cytology, or scan(s)
   D. Physician's **reference to** type of cancer (**histology**) in the medical record
   Note 1: Code the specific histology when documented.
   Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented.

Kidney Histology Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Coding Histology

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

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

   Example 1: Diagnosis for a single tumor is renal cell carcinoma 8312 with the majority or predominant part of tumor being clear cell renal cell carcinoma 8310. Code the subtype/variant: clear cell renal cell carcinoma 8310.

   Example 2: Diagnosis for a single tumor is neuroendocrine tumor 8041 with minority of tumor being large cell neuroendocrine tumor 8013. Code the subtype/variant: large cell neuroendocrine tumor 8013.

   Example 3: Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.

   Note 1: The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.

   Example: When the diagnosis is adenocarcinoma with a clear cell **carcinoma** component, code clear cell carcinoma 8310.

   Negative Example: When the diagnosis is simply adenocarcinoma with a clear cell component, code adenocarcinoma NOS 8140. Do not assume this is a clear cell carcinoma. This could be clear cell differentiation or features.

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

2. Code the histology 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".

   Note: Do not code differentiation or features when there is no specific ICD-O code.

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

      Example: Outpatient biopsy says probably papillary renal cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology papillary renal cell carcinoma. The case meets the criteria in #**3A.**

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

      Example 1: The pathology diagnosis is renal cell carcinoma consistent with chromophobe renal cell carcinoma. The oncology consult says the patient has chromophobe renal cell carcinoma of the right kidney. This is clinical confirmation of the diagnosis, code chromophobe renal cell carcinoma. The case meets the criteria in **bullet 1**.

      Example 2: The pathology diagnosis is neuroendocrine tumor consistent with large cell neuroendocrine tumor. The treatment plan says the patient will receive treatment for large cell neuroendocrine tumor. Treatment plan confirms large cell neuroendocrine tumor; code large cell neuroendocrine tumor. The case meets the criteria in **bullet 2**.

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

Kidney Histology Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

List of Ambiguous Terminology

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

4. **Do not code** histology when described as:

   - Architecture
   - Foci; focus; focal
   - Pattern

Kidney Histology Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule H1 Code the histology when only **one histology** is present.

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

Note 2: When the histology is **not listed** in **Table 1** use the **ICD-O** and all **updates**.

Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 1, ICD-O or all updates.

Rule H2 Code the **NOS** histology when there are:

- A **NOS** and **two or more variants** of that **NOS** present in the tumor **OR**
- **Two or more variants** of a NOS present in the tumor

Example 1: The diagnosis is a single tumor with renal cell carcinoma (RCC) 8312, papillary renal cell carcinoma 8260, and mucinous tubular and spindle cell carcinoma 8480. Papillary renal cell carcinoma and mucinous tubular and spindle cell carcinoma are subtypes/variants of renal cell carcinoma. Code the histology to the NOS, RCC 8312.

Example 2: The diagnosis is spindle cell rhabdomyosarcoma 8912 and alveolar rhabdomyosarcoma 8920. Both are subtypes/variants of rhabdomyosarcoma 8900. Code the NOS, rhabdomyosarcoma.

Informational Item: WHO 4th edition Tumors of the Urinary System has proposed ICD-O code 8323/1 for clear cell papillary renal cell carcinoma. This has not been approved for implementation by the standard setters in 2018.

Note: Use **Table 1** in the Equivalent Terms and Definitions to determine NOS and subtype/variant.

Rule H3 Code the **subtype/variant** when a **NOS** and a **single subtype/variant** of that NOS are present.

- Renal cell carcinoma NOS **8312** and a subtype/variant of RCC
- Rhabdomyosarcoma **8900** and a subtype/variant of rhabdomyosarcoma
- Well differentiated neuroendocrine tumor **8240** and subtype/variant of well differentiated neuroendocrine tumor

Note: Use **Table 1** in the Equivalent Terms and Definitions to determine NOS and subtype/variant.

This is the end of instructions for Single Tumor.

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

Kidney Histology Rules
C649
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multiple Tumors Abstracted as a Single Primary

Note: Multiple tumors **must be a single primary** to use these rules. See the **Multiple Primary Rules** to determine whether these tumors are a single primary.

Rule H4 Code the histology when only **one** histology is present in **all** tumors.

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

Note 2: When the histology is **not listed** in **Table 1** use the **ICD-O** and all **updates**.

Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 1, ICD-O or all updates.

Rule H5 Code the NOS when there are:

- A **NOS** and **two or more variants** of that **NOS p**resent in the tumors **OR**
- **Two or more variants** of a NOS present in the tumors

Example 1: The diagnosis is a single tumor with renal cell carcinoma (RCC) 8312, papillary renal cell carcinoma 8260, and mucinous tubular and spindle cell carcinoma 8480. Papillary renal cell carcinoma and mucinous tubular and spindle cell carcinoma are subtypes/variants of renal cell carcinoma. Code the histology to the NOS: RCC 8312.

Example 2: The diagnosis is spindle cell rhabdomyosarcoma 8912 and alveolar rhabdomyosarcoma 8920. Both are subtypes/variants of rhabdomyosarcoma 8900. Code the NOS: rhabdomyosarcoma.

Informational Item: WHO 4th edition Tumors of the Urinary System has proposed ICD-O code 8323/1 for clear cell papillary renal cell carcinoma. This has not been approved for implementation by the standard setters in 2018.

Note: Use **Table 1** in the Equivalent Terms and Definitions to determine NOS and subtype/variant.

Rule H6 Code the **subtype/variant** when a **NOS** and a **single subtype/variant** of that NOS are present such as the following:

- Renal cell carcinoma **8312** and a subtype/variant of renal cell carcinoma
- Rhabdomyosarcoma **8900** and a subtype/variant of rhabdomyosarcoma
- Well differentiated neuroendocrine tumor **8240** and subtype/variant of well differentiated neuroendocrine tumor

Note: Use **Table 1** in the Equivalent Terms and Definitions to determine NOS and subtype/variant.

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

Code the histology according to the rule that fits the case

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Introduction

Note 1: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Note 2: Cancers **from many primary sites metastasize** to the **lung**. It is important to **rule out metastases** from another organ/site before abstracting a lung primary.

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

Note 4: Multifocal/multiple discrete foci tumors are often present in lepidic adenocarcinoma, minimally invasive adenocarcinoma, and adenocarcinoma in situ; these multiple foci may be referred to as ground-glass/lepidic.

Note 5: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Changes from 2007 MPH Rules

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

Note 1: Changes are **implemented slowly** over time, so it is not unusual for a pathology report to use an obsolete term. **Obsolete** terms and codes **can be used** when they are the **only information** available.

Note 2: WHO 4th Ed Tumors of Lung 2015 has a new classification of adenocarcinoma which is a significant change from the 2004 WHO classification. One of the major changes is discontinuing usage of the term **bronchioloalveolar carcinoma (BAC)** beginning with cases diagnosed 1/1/2018 and forward. The preferred term for BAC is now mucinous adenocarcinoma **8253**.

1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, 2018 Rules instruct "Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection

(two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor)."

2. **New** and **changed** ICD-O histology codes have been added to **Table 3** and are identified by an asterisk. Some of those changes include:
   A. **In situ** and **minimally invasive terms** and codes
   B. **Terms** assigned a **new histology** code
   C. **Histology codes** assigned a **different preferred term** (18 codes with new preferred terms)
3. The following new adenocarcinoma terms and codes have been added. The new terms and codes are **for lung only.** See notes in Table 3.
   A. Mucinous carcinoma/adenocarcinoma
      - **8253/3** when
        - Behavior unknown/not documented (use staging form to determine behavior when available)
        - Invasive
      - **8257/3** when
        - Microinvasive
        - Minimally invasive
      - **8253/2** when
        - Preinvasive
        - In situ

   ***Note*:** Previously, only **invasive** /3 codes were available for mucinous adenocarcinoma of the lung. It has been recognized that not all lung cancers are invasive /3 so new codes were implemented.

   B. Non-mucinous carcinoma/adenocarcinoma
      - **8256/3** when
        - Microinvasive
        - Minimally invasive
      - **8250/2** when
        - Preinvasive
        - In situ

C. Adenocarcinomas (CAP Terminology) Adenocarcinoma, acinar predominant 8551
- Adenocarcinoma, lepidic predominant 8250
- Adenocarcinoma, micropapillary predominant 8265
- Adenocarcinoma, papillary predominant 8260
- Adenocarcinoma, solid predominant 8230

Equivalent or Equal Terms

These terms can be used interchangeably:

- Adenocarcinoma; carcinoma
  - A histology type must be stated for these terms to be equal
  - Example: Acinar carcinoma and acinar adenocarcinoma are both coded 8551
- And; with
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Non-small cell carcinoma **8046**; a broad category which includes all histologies in Table 3 **except** for
  - Small cell carcinoma/neuroendocrine tumors (NET Tumors) **8041** and all subtypes AND
  - Sarcoma NOS **8800** (not a carcinoma) and all subtypes
- Simultaneous; existing at the same time; concurrent; prior to first course treatment
- Site; topography
- Squamous cell carcinoma; SCC; epidermoid carcinoma
- Tumor; mass; tumor mass; lesion; neoplasm; nodule
  - The terms tumor, mass, tumor mass, lesion, neoplasm and nodule are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms **unless** there is a **physician's statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to **determine** multiple **primaries**
  - **Do not** use these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Terms that are NOT Equivalent or Equal

This is a list of terms that are **not equivalent**. There are no casefinding implications.

- **Bilateral** is not equivalent to either **single primary** or **multiple primaries**. See Multiple Primary rules for instructions.
- **Bronchus** is not always equivalent to **mainstem bronchus**. The mainstem bronchus only extends a few centimeters into the lung.
  - Code to mainstem bronchus C340 when it is specifically stated in the operative report and/or documented by a physician
  - When only called bronchus, code to the lobe in which the bronchial tumor is located
- Carcinoma, NOS 8010 is not equivalent to adenocarcinoma, NOS 8140
- **Component** is not equivalent to **type/subtype/variant**
  Note: Component is **only** coded when the pathologist specifies the component as a second **carcinoma.**
- **Lung only: Mucinous** is not equivalent to **colloid**
  Note: The new codes for mucinous adenocarcinoma were implemented so mucinous carcinoma and colloid carcinoma could be analyzed separately.
- **Mucin-producing/mucin-secreting carcinoma 8481** is not equivalent to **mucinous carcinoma 8253** (new code for lung primaries only)
  - Mucin-producing/secreting tumors produce mucin, but not enough to be classified as mucinous carcinoma
  - The terms mucin-producing and mucin-secreting are still reportable. This bullet simply states they are not equivalent to mucinous carcinoma
- **Multilocular** is not equivalent to **multinodular** (see glossary for further information. The electronic glossary will be available in 2019)
- **Phenotype** is not equivalent to **subtype/type/variant**

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Table 1: Coding Primary Site

1. The mainstem bronchus **starts** at the **trachea** and extends only a few centimeters into the lung where it connects with the secondary bronchus and divides into **secondary** bronchi.
   A. Each lobe of the lung has **secondary bronchi**
      i. The **right** lung has **3 secondary bronchi**, one in each of the three lobes: upper; middle, and lower
      ii. The **left** lung has **2 secondary bronchi**, one in each of the two lobes: upper and lower
   B. Code to **mainstem bronchus** C340 when it is **specifically stated** in the operative report and/or documented by a physician.
   C. When **only called bronchus**, code to the lobe in which the bronchial tumor is **located**
2. See the graphic in this document with the endnote "End of Mainstem Bronchus; Start of Terminal/Secondary Bronchus".

Table 1 contains terms used in **physicians' documentation** and on **scans** to describe the location of a tumor.

This table has terms and anatomical descriptions which are not in the ICD-O.

Use this table to determine the **correct site** code. **Do not** use for other fields such as laterality.

Column 1 contains the **terminology used** by **physicians** or on **scans** to **describe lung "masses"** (not lymph nodes).
Column 2 indicates whether the **term** is **used only** for the **right** lung, or only for the **left** lung, or if it is used for **both** the right or left lung.
Column 3 contains the **ICD-O term** and **site code.**

Table begins on next page

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Terminology	Laterality	Site Term and Code
Bronchus intermedius **Carina** **Hilus of lung** **Perihilar**	Bilateral	Mainstem bronchus **C340** *Note*: **Bronchus intermedius** is the portion of the **right mainstem bronchus** between the upper lobar bronchus and the origin of the middle and lower lobar bronchi
Lingula of lung	Left	Upper lobe **C341**
Apex **Apex of lung** **Lung apex** **Pancoast tumor** **Superior lobar bronchus** **Upper lobe bronchi**	Bilateral	Upper lobe **C341**
Middle lobe **Middle lobe bronchi**	Right	Middle lobe **C342**
Base of lung **Lower lobar bronchus** **Lower lobe** **Lower lobe bronchi** **Lower lobe segmental bronchi**	Bilateral	Lower lobe **C343**
Overlapping lesion of lung	Bilateral	Overlapping lesion of lung **C348** *Note*: **One** lesion/tumor which **overlaps two or more** lobes

Table continues on next page

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Terminology	Laterality	Site Term and Code
Bronchus NOS **Bronchogenic** **Extending up to the hilum** **Extending down to the hilar region** **Infrahilar NOS** **Lung NOS** **Pulmonary NOS** **Suprahilar NOS**	Bilateral	Lung NOS **C349** ***Note***: Includes • Multiple tumors in different lobes of ipsilateral lung **OR** • Multiple tumors in ipsilateral lung; unknown if same lobe or different lobe **OR** • Tumor in bronchus, unknown if mainstem or lobar bronchus **OR** • Tumor present, unknown which lobe
Lobar bronchi NOS **Lobar bronchus NOS**	Bilateral	Code the lobe in which the lobar bronchus tumor is present **C34__** ***Note***: When **lobe** of origin is **not documented/unknown**, **code** to lung NOS **C349**

Lung Equivalent Terms and Definitions
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Table 2: Combination/Mixed Histology Codes

Instructions:

1. Compare the **terms** in the **diagnosis** (pathology, cytology, radiographic, clinical) to the terms in **Column 1.**
2. When the terms **match**, use the **combination code** listed in **Column 2.**
3. The **last row** in the table is a **"last resort"** code: adenocarcinoma mixed subtypes 8255.

Note 1: **Do not** use Table 2 in the following situations:

- For tumors with both **invasive** and **in situ** behavior. The **Histology Rules** instruct to code the invasive histology.
- When one of the histologies is described as **differentiation or features.** A histology with differentiation or features is a single histology.
- When the terms are a **NOS** and a **subtype/variant** of that NOS. See the **Histology Rules** for instructions on coding a NOS and a subtype/variant in a single tumor or multiple tumors abstracted as a single primary.

Note 2: Some combinations can be either in situ or invasive; others are limited to a /2 or /3 behavior code.

- When a code is **limited to in situ, /2** will be **added** to the code (both components are in situ)
- When a code is **limited to invasive, /3** will be **added** to the code (both components are invasive)

Note 3: This table is not a complete listing of histology combinations.

Column 1 lists the **required terms** for **the combination code.**
Column 2 lists the **combination term** and **code** for histologies **in Column 1.**

Table begins on next page.

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Required Terms	Combination Histologies and Code
Adenocarcinoma NOS **AND** Squamous cell carcinoma NOS ***Note*:** Cases diagnosed prior to 1/1/2023: Diagnosis **must be** adenocarcinoma NOS and squamous cell carcinoma NOS, **NOT** any of the **subtypes/variants** of adenocarcinoma or squamous cell carcinoma Cases diagnosed 1/1/2023 forward: Subtypes/variants of adenocarcinoma, NOS and keratinizing and/or non-keratinizing variants of squamous cell carcinoma, NOS can be coded adenosquamous carcinoma	Adenosquamous carcinoma **8560**
Giant cell carcinoma **AND** Spindle cell carcinoma ***Note:*** Sarcomatoid carcinoma is not in the histology table because sarcomatoid tumors primarily originate in the mediastinum. The combination code is added for the rare occasion when a tumor occurs within the lung.	Sarcomatoid carcinoma **8033** ***Note:*** Both giant cell carcinoma and spindle cell carcinoma are components of sarcomatoid carcinoma. The most accurate code for a combination of giant cell and spindle cell carcinoma is sarcomatoid carcinoma
Epithelial carcinoma **AND** Myoepithelial carcinoma	Epithelial-myoepithelial carcinoma **8562**

Lung Equivalent Terms and Definitions
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Required Terms	Combination Histologies and Code
Large cell neuroendocrine carcinoma **AND** Adenocarcinoma NOS **OR** Squamous cell carcinoma NOS **OR** Spindle cell carcinoma **OR** Giant cell carcinoma	Combined large cell neuroendocrine carcinoma **8013**
Mucinous carcinoma, invasive **AND** Non-mucinous carcinoma, invasive	Mixed invasive mucinous and non-mucinous carcinoma **8254/3***
Small cell carcinoma/neuroendocrine tumor (NET) ***Note:*** **Includes subtypes/variants** of small cell/neuroendocrine tumor. See Table 3 for subtypes/variants. **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**

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Required Terms	Combination Histologies and Code
Squamous cell carcinoma (epidermoid carcinoma) **AND** Large cell non-keratinizing squamous cell carcinoma ***Note:*** Squamous cell carcinoma and epidermoid carcinoma are synonyms	Squamous cell carcinoma, large cell, nonkeratinizing **8072**
Squamous cell carcinoma (epidermoid carcinoma) **AND** Small cell nonkeratinizing squamous cell carcinoma ***Note:*** Squamous cell carcinoma and epidermoid carcinoma are synonyms	Squamous cell carcinoma, small cell, nonkeratinizing **8073**
Squamous cell carcinoma, keratinizing **AND** Squamous cell carcinoma, non-keratinizing	Squamous cell carcinoma, NOS **8070**
Squamous cell (epidermoid) carcinoma **AND** **One or both** of the following: • Sarcomatoid carcinoma • Spindle cell carcinoma ***Note 1:*** **Does not include subtypes/variants** of squamous cell. See **Table 3** for subtypes/variants. ***Note 2:*** Squamous cell carcinoma and epidermoid carcinoma are synonyms.	Squamous cell carcinoma, sarcomatoid **8074** Squamous cell carcinoma, spindle cell **8074**

Lung Equivalent Terms and Definitions
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Required Terms	Combination Histologies and Code
Diagnosis must be a single tumor which meets one of the following two criteria: 1. <u>At least two</u> of the subtypes/variants of adenocarcinoma **AND** <u>percentages of each type are unknown/not stated **OR** they are equal percentages</u> • Acinar adenocarcinoma • Clear cell adenocarcinoma • Lepidic adenocarcinoma ***Note***: Lepidic adenocarcinoma may or may not have mucinous components. • Micropapillary adenocarcinoma • Papillary adenocarcinoma • Solid adenocarcinoma • Well-differentiated fetal adenocarcinoma ***Note:*** This includes a diagnosis of adenocarcinoma **AND** at least two subtypes/variants of adenocarcinoma. 2. A combination of histologies **<u>not listed on previous rows</u>** of this table.	Adenocarcinoma with mixed subtypes **8255/3** ***Note 1***: 8255 is a "last resort" code. ***Note 2:*** See the **Histology Rules** to determine when it is appropriate to use this code for combination histologies other than adenocarcinoma subtypes/variants. ***Note 3:*** 8255 does not apply to squamous cell carcinoma NOS and/or subtype/variants of SCC.

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

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

Use Table 3 as directed by the **Histology Rules** to assign the more common histology codes for lung tumors.

Note 1: Rare histologies may not be listed in the table. When a histology term is not found, reference ICD-O and all updates.
Note 2: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 3, ICD-O or ICD-O updates.
Note 3: Behavior codes are listed when the term has only one possible behavior (either a /2 or /3). For histologies which may be either /2 or /3, a behavior code is not listed. Code behavior as documented in the pathology report.
Note 4: Only use the histology code from the table when the diagnosis is **EXACTLY** the term listed.
Note 5: Sarcomatoid carcinoma is most frequently a tumor of the mediastinum, so it is not listed in this table.

IMPORTANT NOTE 1: Non-small cell lung carcinoma (NSCLC) is a broad group of cancers which includes all **carcinoma types** in Table 3 with the **exception** of:

- Neuroendocrine carcinoma (NEC), neuroendocrine tumors (NET) AND
  - Small cell carcinoma/neuroendocrine tumors/all subtypes of small cell carcinoma AND
  - Large cell neuroendocrine carcinoma/combined large cell neuroendocrine carcinoma AND
- Sarcoma NOS 8800 (not a carcinoma) **AND**
  - All subtypes of sarcoma NOS

NSCLC is usually adenocarcinoma, squamous cell carcinoma, or large-cell carcinoma. See the instructions for coding histology when NSCLC is the diagnosis.

IMPORTANT NOTE 2: The small cell neuroendocrine carcinoma row has been deleted in the 2024 update and replaced with new rows for neuroendocrine carcinoma (NEC) and neuroendocrine tumor (NET). This change is based on the 5th Ed WHO Classification of Lung tumors book and current concepts.

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants**

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

Table begins on next page

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Adenocarcinoma 8140 **Note 1:** Mucinous adenocarcinoma for lung only is coded as follows: • **8253/3*** when o Behavior unknown/not documented (use staging form to determine behavior when available) o Invasive • **8257/3*** when o Microinvasive o Minimally invasive • **8253/2*** when o Preinvasive o In situ **Note 2:** Non-mucinous adenocarcinoma for lung only is coded as follows: • **8256/3*** when o Microinvasive o Minimally invasive • **8250/2*** when o Preinvasive o In situ **Note 3**: The term "mucinous carcinoma/adenocarcinoma NOS" is not recommended for lung; specific mucinous terms should be used. If a diagnosis states mucinous carcinoma/adenocarcinoma, NOS and a subtype/variant, code the subtype/variant.	Adenocarcinoma NOS Adenocarcinoma in situ **8140/2** Adenocarcinoma invasive **8140/3** Adenocarcinoma, non-mucinous NOS Invasive non-mucinous adenocarcinoma **8140/3** Minimally invasive adenocarcinoma NOS **8140/3**	Acinar adenocarcinoma/adenocarcinoma, acinar predominant **(for lung only) 8551*** Adenoid cystic/adenocystic carcinoma **8200** Colloid adenocarcinoma **8480** Enteric adenocarcinoma/pulmonary intestinal-type adenocarcinoma **8144** Fetal adenocarcinoma **8333** Lepidic adenocarcinoma/adenocarcinoma, lepidic predominant **8250/3*** Mucinous carcinoma/adenocarcinoma **(for lung only; See Note 3)** in situ **8253/2*;** invasive **8253/3*** minimally invasive **8257/3*** microinvasive **8257/3*** preinvasive **8253/2*** Micropapillary adenocarcinoma/adenocarcinoma, micropapillary predominant **8265** Mixed invasive mucinous and non-mucinous adenocarcinoma **8254*** Non-mucinous adenocarcinoma **(for lung only)** in situ **8250/2*** microinvasive **8256/3*** minimally invasive **8256/3*** preinvasive **8250/2*** Papillary adenocarcinoma/adenocarcinoma, papillary predominant **8260** Solid adenocarcinoma/adenocarcinoma, solid predominant **8230**

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Adenosquamous carcinoma 8560		
Carcinosarcoma 8980/3		
Diffuse pulmonary lymphangiomatosis 9170/3 *Note*: Diffuse pulmonary lymphangiomatosis is a diffuse proliferation of lymphatic channels and smooth muscle along otherwise normal lymphatic vessels of lungs, pleura, and mediastinum. Primarily occurs in infants and children.		
Epithelial-myoepithelial carcinoma 8562 *Note:* Adenomyoepithelioma, epithelial/myoepithelial tumor of unproven malignant potential were thought to be adenomas (not reportable) prior to 2018. These histologies are now designated as low-grade carcinomas based on lymph node metastasis, local invasion, and aggressiveness	Adenomyoepithelioma* Epimyoepithelial carcinoma Epithelial-myoepithelial tumor of unproven malignant potential* Malignant mixed tumor comprising epithelial and myoepithelial cells Pneumocytic adenomyoepithelioma*	
Epithelioid hemangioepithelioma 9133		
Giant cell carcinoma 8031		
Hyalinizing clear cell carcinoma 8310		

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Intrapulmonary thymoma (arising within lung) 8580/3 *Note:* **Intrapulmonary** thymoma is **always malignant** /3.		
Large cell carcinoma 8012 *Note 1:* A diagnosis of large cell carcinoma is usually followed by further diagnostic testing to identify the subtype/variant. *Note 2:* The diagnosis of large cell carcinoma usually happens when there is a small amount of tissue (FNA), cytology, or when the tumor is highly differentiated. Large cell carcinoma lacks the features of small cell carcinoma, adenocarcinoma, or squamous carcinoma. *Note 3:* Large cell carcinoma with **neuroendocrine (NE) differentiation** lacks NE morphology and is coded as **large cell** carcinoma, **not large cell neuroendocrine** carcinoma.	Large cell anaplastic carcinoma Large cell carcinoma NOS Large cell carcinoma with no additional stains (subtype/variant – no ICD-O code) Large cell carcinoma with null immunohistochemical features (subtype/variant – no ICD-O code) Large cell carcinoma with unclear immunohistochemical features (subtype/variant – no ICD-O code) Large cell undifferentiated carcinoma	

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Lymphoangioleiomyomatosis 9174/3 Note: Locally destructive mesenchymal neoplasm.		
Large cell neuroendocrine carcinoma 8013 **Note:** Per WHO, both large cell neuroendocrine carcinoma, NOS and combined large cell neuroendocrine carcinoma are coded 8013. See Table 2 for histologies included in combined large cell neuroendocrine carcinoma	Combined large cell neuroendocrine carcinoma	
Lymphoepithelioma-like carcinoma 8082		
Melanoma 8720		
Mucoepidermoid carcinoma 8430 ***Note:*** As of 1/1/2023, mucoepidermoid tumor **is no longer a** synonym of mucoepidermoid carcinoma in WHO		
Myoepithelial carcinoma 8982		
Neuroendocrine carcinoma (NEC) 8246		Combined small cell carcinoma **8045** Small cell carcinoma/small cell neuroendocrine carcinoma **8041**

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Neuroendocrine tumor, NOS (NET) 8240	Bronchial adenoma, carcinoid Carcinoid, NOS Carcinoid tumor, NOS Neuroendocrine tumor, grade 1 Neuroendocrine tumor, low grade Neuroendocrine carcinoma, well differentiated Typical carcinoma	Atypical carcinoid/Neuroendocrine tumor, grade 2 Neuroendocrine tumor, grade 3/Neuroendocrine carcinoma, moderately differentiated **8249**
NUT carcinoma 8023/3* **NUT: nuclear protein in tests** **NUT/M1 gene rearrangement**	Aggressive t(15:19) positive carcinoma BET-rearranged carcinoma Carcinoma with t(15:19) translocation Midline carcinoma of children and young adults with NUT rearrangement Midline lethal carcinoma NUT midline carcinoma	
PEComa malignant 8714/3 ***Note:*** Tumor displays perivascular epithelioid (PEC) differentiation	PEComa of the lung PEComa, malignant	

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Pleomorphic carcinoma 8022 *Note 1*: The **definition** of pleomorphic carcinoma is that it is a **subtype** of **sarcomatoid** carcinoma. It has at least 10% spindle or giant cells. *Note 2*: Pleomorphic carcinoma has components of **adenocarcinoma** and/or **large cell** carcinoma, also **squamous** carcinoma.		
Pleuropulmonary blastoma 8973/3 *Note*: Pleuropulmonary blastoma is an embroynal tumor and differs from pulmonary blastoma		
Pulmonary blastoma 8972/3 ***Note:*** Pulmonary blastoma is a biphasic tumor that consists of low-grade/WD fetal adenocarcinoma and primitive mesenchymal differentiation (osteosarcoma, chondrosarcoma, or rhabdomyosarcoma).		
Sarcoma NOS 8800/3		Biphasic synovial sarcoma **9043/3** Epithelioid cell synovial sarcoma **9042/3** Pulmonary artery intimal sarcoma/low-grade malignant myxoid endobronchial tumor **9137/3** Pulmonary myxoid sarcoma with EWSR1 -CREB1 translocation **8842/3** Spindle cell synovial sarcoma **9041/3** Synovial sarcoma **9040/3**
Spindle cell carcinoma 8032		

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Squamous cell carcinoma 8070	Epidermoid carcinoma Epidermoid carcinoma NOS Squamous carcinoma Squamous cell carcinoma NOS Squamous cell epithelioma Squamous cell carcinoma in situ **8070/2**	Basaloid carcinoma/basaloid squamous cell carcinoma **8083** Keratinizing squamous cell carcinoma **8071** Non-keratinizing carcinoma **8072**
Thoracic SMARCA4-deficient undifferentiated tumor 8044/3		

***New codes/terms approved by IARC/WHO Committee for ICD-O.**

Lung Equivalent Terms and Definitions
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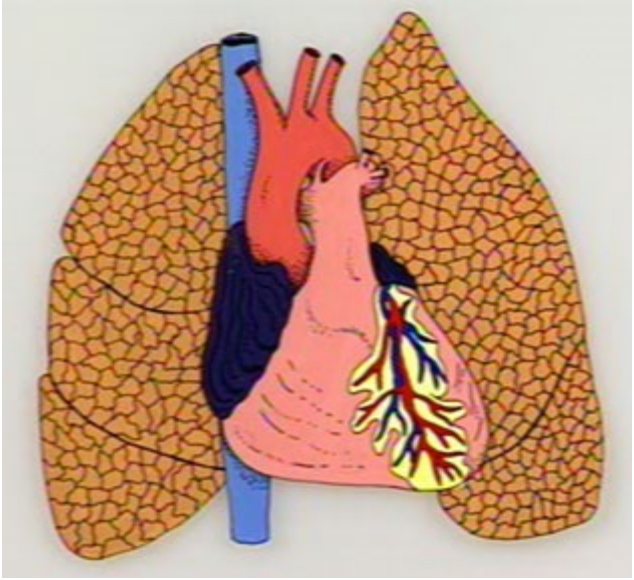
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Illustrations



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Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Mediastinum
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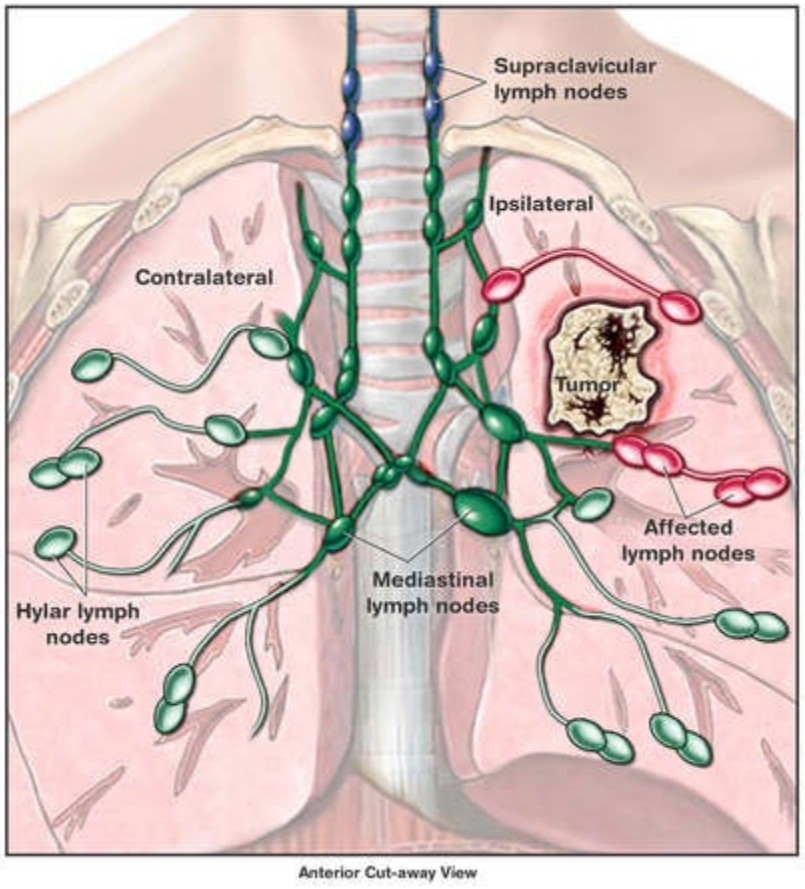

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Lymph Nodes Lung
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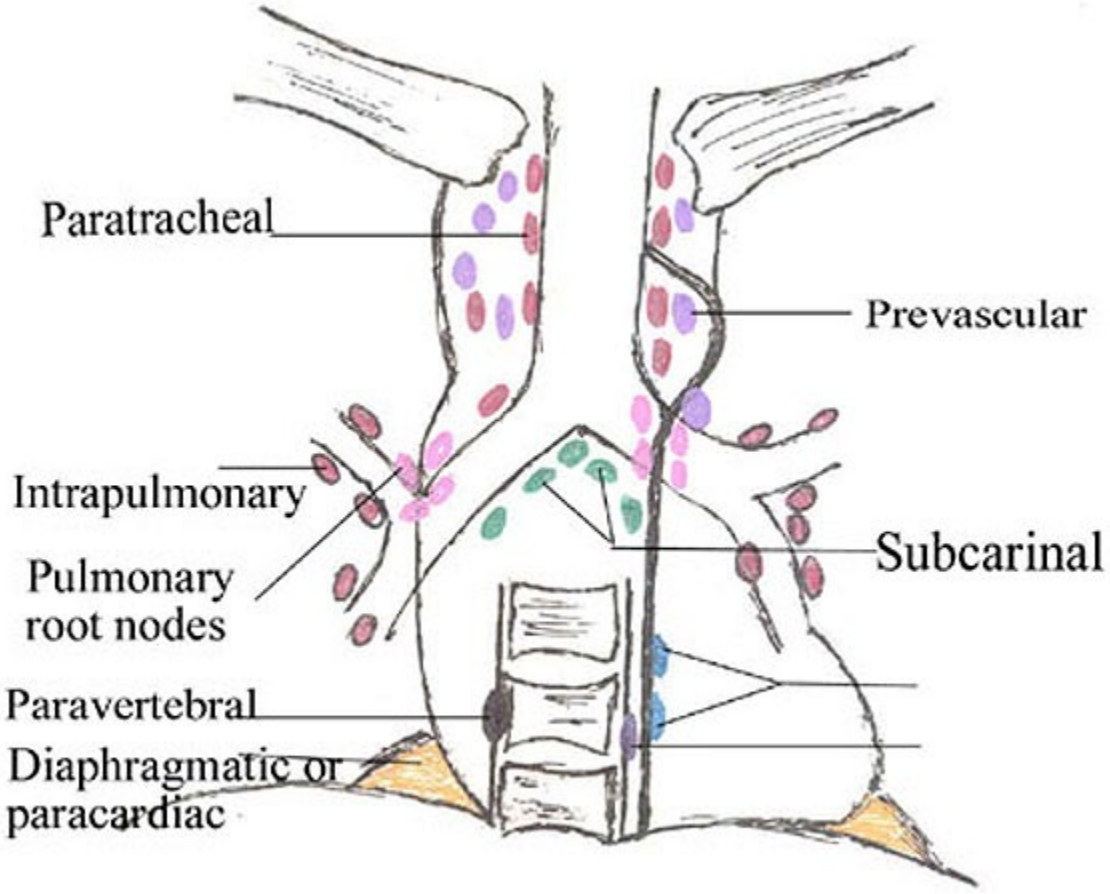

Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Inside the Lung
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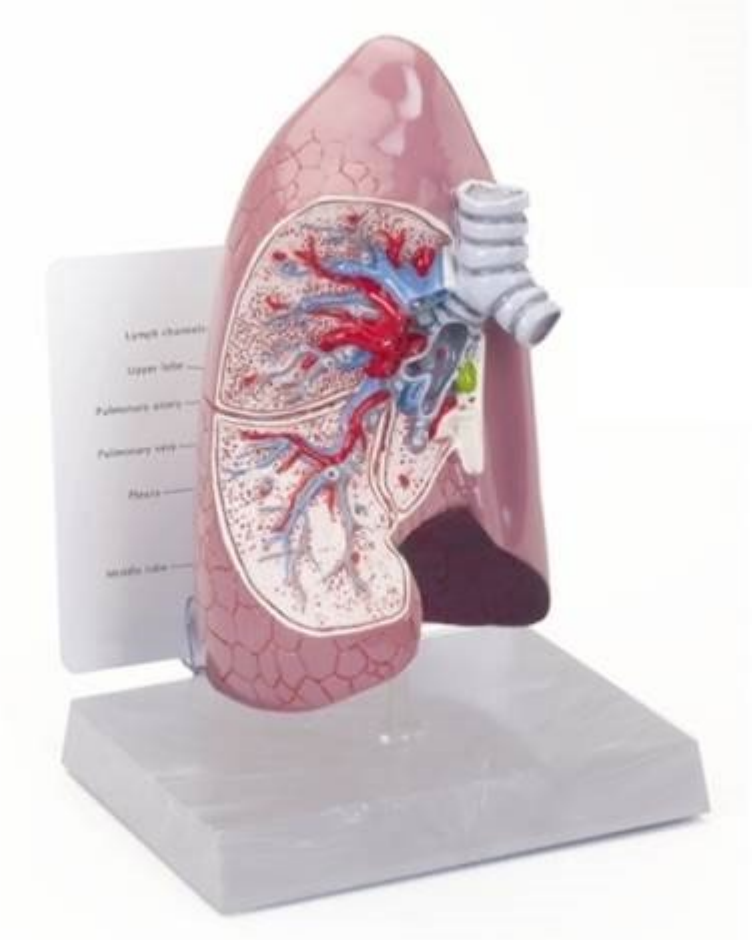

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Gross Anatomy of Lung
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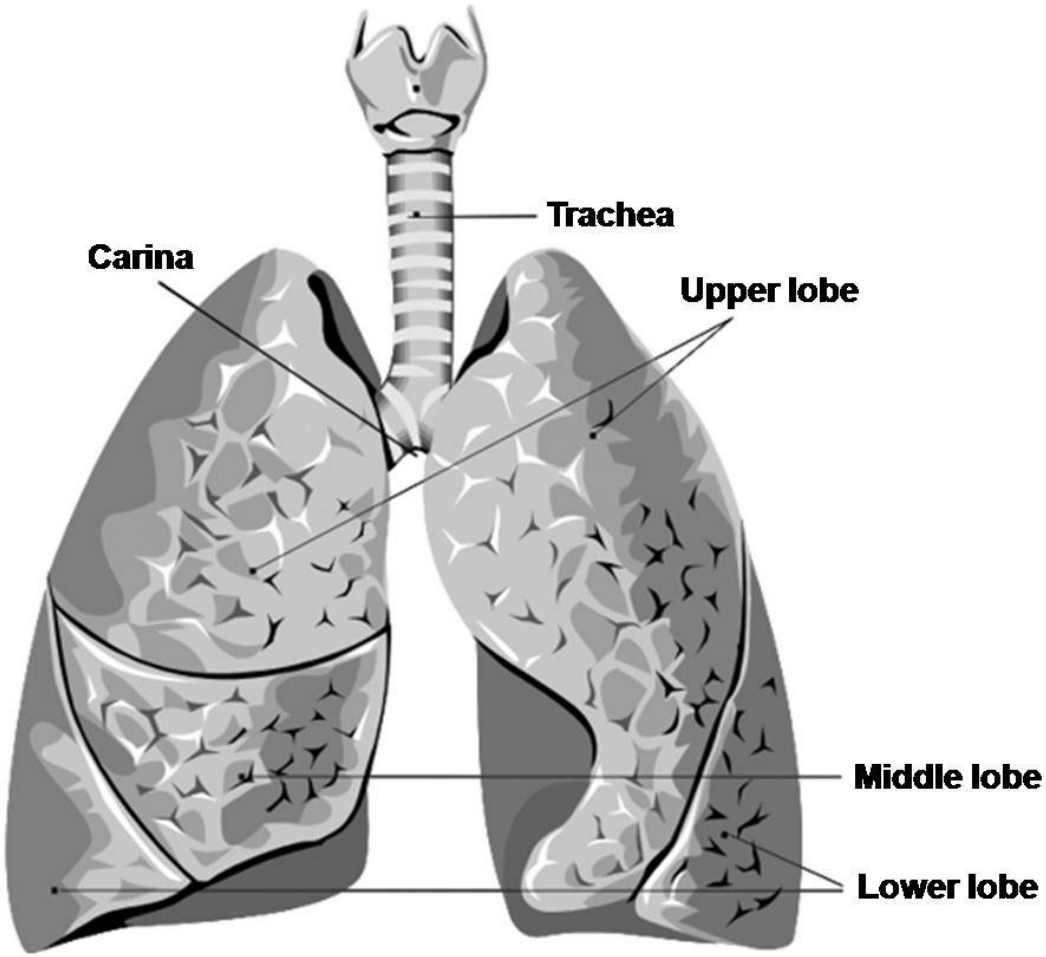

Lung Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

End of Mainstem Bronchus; Start of Terminal/Secondary Bronchus
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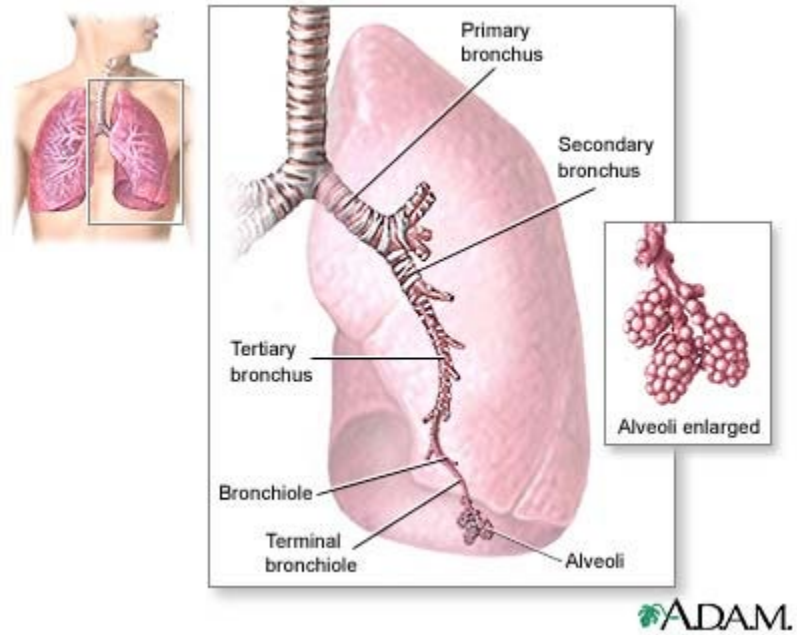

Lung Multiple Primary Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Adrenal glands
- Bone
- Brain
- Discontinuous lesions in adjacent/contiguous organs
- Discontinuous lesions in chest wall
- Discontinuous lesions/nodules in soft tissue adjacent to primary site
- Regional or distant lymph nodes as identified in Summary Staging Manual
- Esophagus
- Heart
- Liver
- Trachea

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors

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

Lung Multiple Primary Rules
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(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule M2 Abstract a **single primary**[i] when there is a **single tumor.**
Note 1: A single tumor is <u>always</u> a single primary.
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor

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

Multiple Tumors

Note: Multiple tumors may be a single primary or multiple primaries.

Rule M3 Abstract **multiple primaries**[ii] when there are **separate, non-contiguous** tumors in sites with ICD-O **site** codes that differ at the second C**<u>X</u>**xx and/or third character Cx**<u>X</u>**x.
Note: When **codes differ** at the second or third characters, the tumors are in **different** primary **sites.**

Rule M4 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being **clinically disease-free** for greater than **three years** after the original diagnosis or last recurrence.
Note 1: **Clinically** disease-free means that there was **no <u>evidence</u>** of recurrence in the same lung on follow-up.
- Scans are NED

Note 2: When there is a recurrence less than or equal to three years of diagnosis, the **"clock"** starts over. The time interval is calculated from the **date of last recurrence.** In other words, the patient must have been disease-free for greater than three years from the date of the last recurrence.
Note 3: When it is **unknown/not documented** whether the patient had a recurrence, use **date of diagnosis** to compute the time interval.
Note 4: The physician may state this is a **recurrence**, meaning the patient had a previous lung tumor and now has another lung site tumor. **Follow the rules**; do not attempt to interpret the physician's statement.

Lung Multiple Primary Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M5 Abstract **multiple primaries**[ii] when there is **at least one** tumor that is **small cell** carcinoma **8041** or any small cell subtypes/variants and another tumor that is **non-small cell** carcinoma **8046** or any non-small cell carcinoma subtypes/variants.

Note 1: Small cell carcinoma and non-small cell carcinoma are the two major classifications/divisions for lung cancer.

- See **Table 3** in Equivalent Terms and Definitions for terms and codes for small cell carcinoma and all of the subtypes/variants
- With the exception of small cell/neuroendocrine carcinoma and sarcomas, **all other histologies** listed in **Table 3** in Equivalent Terms and Definitions are **non-small cell** carcinoma

Note 2: It is **irrelevant** whether the tumors are in the **ipsilateral** (same) lung or are **bilateral** (both lungs).

Rule M6 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3, **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

Note 1: The tumors may be subtypes/variants of the **same** or **different** NOS histologies.

- **Same NOS:** Colloid adenocarcinoma 8480/3 and lepidic adenocarcinoma 8250/3 are both subtypes of adenocarcinoma NOS 8140/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS:** Keratinizing squamous cell carcinoma 8071/3 is a subtype of squamous cell carcinoma NOS 8070; Lepidic adenocarcinoma 8250/3 is a subtype of adenocarcinoma 8140/3. They are distinctly different histologies. Abstract multiple primaries.

Note 2: The tumors may be different **behaviors:** Acinar adenocarcinoma 8551/3 and mucinous carcinoma, in situ 8253/2 are both subtypes of adenocarcinoma NOS 8140/3 but are distinctly different histologies. Abstract multiple primaries.

Rule M7 Abstract a **single primary**[i] when synchronous, separate/non-contiguous tumors **in the same lung** are on **the same row** in **Table 3** in the Equivalent Terms and Definitions.

Note 1: Tumors must be **in the same lung.**

Note 2: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3)

Lung Multiple Primary Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M8 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are:

- On different rows in **Table 3** in the Equivalent Terms and Definitions
- A combination code in **Table 2** and a code from **Table 3**

Note 1: Timing is irrelevant. Tumors may be synchronous or non-synchronous.

Note 2: Each row in the table is a distinctly different histology.

Example 1: In 2018, the patient has non-mucinous adenocarcinoma **8140/3**. Patient returns in 2019 with large cell carcinoma **8012/3**. These histologies are on different rows in Table 3. Abstract two primaries.

Example 2: In 2017, patient had epithelial-myoepithelial carcinoma **8562** (combination code from Table 2). In 2020, the patient returned with a myoepithelial carcinoma **8982** in the same lung (histology from Table 3). Abstract two primaries.

Rule M9 Abstract a **single primary**[i] when there are **simultaneous multiple** tumors:

- In **both** lungs (multiple in right and multiple in left) **OR**
- In the **same** lung **OR**
- **Single** tumor in one lung; **multiple** tumors in **contralateral** lung

Note 1: Tumors may be combinations of:

- In situ and invasive **OR**
- NOS and subtype/variant (See **Table 3** in the Equivalent Terms and Definitions)
- Cancer NOS **8000** or carcinoma NOS **8010** and any other histology

Note 2: Examples of NOS and subtypes/variants include:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Squamous cell carcinoma **8070** and a subtype/variant of squamous cell carcinoma
- NSCLC **8046** and a subtype/variant of NSCLC
- Carcinoma NOS **8010** and adenocarcinoma

Note 3: Code multiple primaries only when there is **proof** that one of the tumors is a different histology. Proof is any one of the following:

- Pathology from a biopsy or resection proves tumors are different histologies
- Attending, oncologist, or pulmonologist state unequivocally that the tumors are different primaries
  - **Unequivocal** means that **no words** such as "**probable"** are used in the statement. Terms which are on the "ambiguous terms" list such as "probable" cannot be used to prove different primaries.

Note 4: When there are multiple tumors in one or both lungs, the physician usually biopsies only one mass/tumor. They treat the patient based on that single biopsy, assuming all of the masses/tumors are the same histology.

Note 5: Multiple tumors in the same lung, or both lungs, or single tumor in one lung and multiple tumors in the contralateral lung must be diagnosed **simultaneously (same time) to apply this rule. Refer to the rules when multiple tumors are not diagnosed simultaneously.**

Rule M10 Abstract a **single primary**[i] when an **in situ** tumor is diagnosed **after** an **invasive** tumor **AND** tumors occur in the same lung.

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.

Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS. See **Table 3** in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

Note 3: The **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.

Rule M11 Abstract **multiple primaries**[ii] when there is a **single** tumor in **each lung** (one tumor in the right lung and one tumor in the left lung).

Note 1: The only **exception** is when there is **proof** that one tumor is **metastatic**. Proof is any one of the following:

- Tissue from both tumors is compared and the pathologic diagnoses definitively says one tumor is metastatic
- Attending physician, oncologist, or pulmonologist state unequivocally that the tumor in the contralateral lung is metastatic
    - Unequivocal means that no words such as "probably possibly, most likely, etc." are used in the statement. Terms which are on the "ambiguous terms" list make the statement equivocal (cannot be used to prove metastases)

Note 2: Lung **metastases usually** present as multiple tumors/masses. A single tumor in each lung is unlikely to be a single primary (e.g. metastatic to the contralateral lung).

Note 3: The term "**bilateral**" **is not** a synonym for a **single primary**. It is simply a statement that there are tumors in both lungs.

Note 4: This rule is based on **long-term epidemiologic studies** of multiple primaries. The specialty medical experts (SME) and the CoC site physician teams reviewed and approved these rules. Many of the CoC site team physicians were also authors, co-authors, or editors of the AJCC Staging Manual.

Note 5: Lymph node involvement is recorded in staging criteria.

Note 6: Tumors do **not** need to be diagnosed at the same time (do not need to be simultaneous or synchronous).

Lung Multiple Primary Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M12 Abstract a **single primary**[i] (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60** days **after** an **in situ** tumor in the same lung.

Note 1: The rules are **hierarchical.** Only use this rule when none of the previous rules apply.
Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS.
Note 3: When the case has been abstracted, **change behavior** code on original abstract from /2 to /3.
Note 4: Do **not** change **date of diagnosis**.
Note 5: If the case has already been submitted to the central registry, **report** all changes.
Note 6: The physician may **stage both** tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).
Note 7: See the **CoC** and **SEER** manuals for **instructions** on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M13 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor in the same lung.

Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: Abstract **both** the invasive and in situ tumors.
Note 3: Abstract as multiple primaries even if **physician states** the invasive tumor is disease **recurrence** or **progression.**
Note 4: This rule is based on long-term epidemiologic studies of recurrence intervals. The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were also authors, co-authors, or editors of the AJCC Staging Manual.

Rule M14 Abstract a **single primary**[i] when none of the previous rules apply.

Note: Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

This is the end of instructions for Multiple Tumors

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[i] Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.
[ii] Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

Lung Histology Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note: WHO 4th Ed Tumors of Lung: in 2011 has a new classification of adenocarcinoma which is a significant changes from the 2004 WHO classification. One of the major changes is discontinuing usage of the term bronchioloalveolar carcinoma (BAC) beginning with cases diagnosed 1/1/2018 and forward. The preferred term for BAC is now mucinous adenocarcinoma **8253**.

Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

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

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

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

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

Code the **most specific** histology from either **resection** or **biopsy**.
Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

This is a hierarchical list of source documentation.

1. **Tissue or pathology** report from primary site (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP

C. CAP protocol

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

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

Note 3: The CAP protocol is a checklist which:

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

2. **Cytology** (Fine needle biopsy from primary site, pleural fluid or pericardial fluid)

Example: Fine needle aspiration shows squamous cell carcinoma and the resection pathology shows invasive adenocarcinoma. Code adenocarcinoma 8140/3.

3. Tissue/pathology from a **metastatic** site

Note 1: Code the behavior /3.

Note 2: The **tissue** from a **metastatic s**ite often shows **variations** from the primary tumor. When it is the only tissue available, it is **more accurate** than a **scan**.

4. **Scan:** The following list is **in priority** order.
   A. CT
   B. PET
   C. MRI
   D. Chest X-ray

5. Code the histology **documented** by the physician when none of the above are available. Use the **documentation** in the following **priority order:**
   A. Treatment Plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that **refers to original pathology, cytology, or scan(s)**
   D. Physician's **reference to** type of cancer (**histology**) in the medical record

Note 1: Code the specific histology when documented.

Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented.

Lung Histology Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Coding Histology

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

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

   Example 1: Diagnosis for a single tumor is adenocarcinoma 8140 with the majority or predominant part of tumor being acinar adenocarcinoma 8551. Code the subtype/variant: acinar adenocarcinoma 8551.

   Example 2: Diagnosis for a single tumor is squamous cell carcinoma 8070 with minority of tumor being keratinizing squamous cell carcinoma 8071. Code the subtype/variant: keratinizing squamous cell carcinoma 8071.

   Example 3: Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.

   Note 1: The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.

   Example: When the diagnosis is adenocarcinoma with a component of medullary **carcinoma**, code medullary carcinoma 8510.

   Negative Example: When the diagnosis is simply adenocarcinoma with a medullary component, code adenocarcinoma NOS 8140. Do not assume this is a medullary carcinoma. This could be medullary differentiation or features.

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

2. **Code** the histology 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".
   Note: Do not code differentiation or features when there is no specific ICD-O code.

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

      Example: Outpatient biopsy says probably squamous cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology squamous cell carcinoma. The case meets the criteria in #**3A.**

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

      Example: The pathology diagnosis is NSCLC consistent with adenocarcinoma. The oncology consult says the patient has adenocarcinoma of the right lung. This is clinical confirmation of the diagnosis, code adenocarcinoma. The case meets the criteria in **bullet 1.**

      Example: The pathology diagnosis is NSCLC consistent with squamous cell carcinoma. The treatment plan says the patient will receive treatment for squamous cell carcinoma. Treatment plan confirms squamous cell carcinoma; code squamous cell carcinoma. The case meets the criteria in **bullet 2.**

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

List of Ambiguous Terminology

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

4. **DO NOT CODE** histology described as:
   - Architecture
   - Foci; focus; focal
   - Pattern (**Exception**: See Rule H7)

Single Tumor

Rule H1 Code **mucinous** adenocarcinoma as follows (for lung only):

- **8253/3** when
  - Behavior unknown/not documented (use staging form to determine behavior when available)
  - Invasive
- **8257/3** when
  - Microinvasive
  - Minimally invasive
- **8253/2** when
  - Preinvasive
  - In situ

Note 1: When mucinous carcinoma is mixed with another histology, such as adenocarcinoma and mucinous carcinoma, code mucinous **ONLY** when mucinous is **documented** to be **greater than 50%** of the tumor.

Note 2: These **new codes and terms** will allow mucinous adenocarcinoma to be analyzed separately from colloid carcinoma.

Note 3: Changes take place over time. **Pathologists may not use** terms "minimally invasive" and "pre-invasive" **immediately**. Code the pathology diagnosis.

Rule H2 Code **non-mucinous** adenocarcinoma as follows:

- **8256/3** when
  - Microinvasive
  - Minimally invasive
- **8250/2** when
  - Preinvasive
  - In situ

Note 1: These are new codes and terms.
Note 2: Pathologists may not use the terms "minimally invasive" and "pre-invasive" immediately. Code the pathology diagnosis.

Rule H3 Code the specific histology when the diagnosis is **non-small cell lung carcinoma** (NSCLC) **consistent with** (or any other ambiguous term) **a specific carcinoma** (such as adenocarcinoma, squamous cell carcinoma, etc.) when:

- The histology is clinically confirmed by a physician (attending, pathologist, oncologist, pulmonologist, etc.)
- The patient is treated for the histology described by an ambiguous term

Note 1: If the case does not meet the criteria in the first two bullets, code non-small cell lung cancer (NSCLC) 8046.
Note 2: If the case is accessioned (added to your database) based on a **single histology** described by ambiguous terminology and no other histology information is available/documented, then code that histology.

Example 1: The pathology diagnosis is NSCLC consistent with adenocarcinoma. The oncology consult says the patient has adenocarcinoma of the right lung. This is clinical confirmation of the diagnosis, code adenocarcinoma. The case meets the criteria in **bullet 1**.

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

Example 3: Outpatient biopsy says probably squamous cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology squamous cell carcinoma. The case meets the criteria in **Note 2**.

Lung Histology Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule H4 Code the histology when only **one histology** is present.

Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in **Table 3** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Table 3,** use the **ICD-O** and all **updates**.

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

Note 4: This includes coding non-small cell carcinoma when it is the only diagnosis available.

Rule H5 Code the **invasive** histology **when in situ** and **invasive** histologies are present.

Note 1: Histologies may be **NOS** and a **subtype/variant**.

Note 2: When the NOS is invasive and the subtype/variant is situ, code the NOS (invasive).

Example: The histologies are mucinous adenocarcinoma in situ **8253/2** and invasive adenocarcinoma NOS **8140/3.** Code the invasive histology: adenocarcinoma **8140/3**.

Rule H6 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS, such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Mucinous adenocarcinoma and a subtype/variant of mucinous adenocarcinoma
- Non-small cell carcinoma **8046** and a subtype/variant of non-small cell carcinoma
- Sarcoma **8800** and a subtype/variant of sarcoma
- Neuroendocrine carcinoma (NEC) **8246** and a subtype/variant of NEC
- Neuroendocrine tumor (NET) **8240** and a subtype/variant of NET
- Squamous cell carcinoma **8070** and a subtype/variant of squamous cell carcinoma

Note: See **Table 3** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

Rule H7 Code the histology that comprises the **greatest percentage** of tumor when two or more of the following histologies are present:

- Acinar adenocarcinoma / Adenocarcinoma, acinar predominant **8551**
- Lepidic adenocarcinoma / Adenocarcinoma, lepidic predominant **8250**
- Micropapillary adenocarcinoma / Adenocarcinoma, micropapillary predominant **8265**
- Papillary adenocarcinoma / Adenocarcinoma, papillary predominant **8260**
- Solid adenocarcinoma / Adenocarcinoma, solid predominant **8230**

Note 1: The rules are hierarchical, so the tumors are **NOT** a NOS and subtype/variant.

Note 2: If the percentages are unknown/not documented, or are equal percentages, continue through the rules.

Note 3: CAP Lung Protocol now allows pathologists to identify the bulleted histologies as **pattern** along with percentages. The histology pattern with the greatest percentage can be coded. This is an **exception** to the histology coding instruction to not code pattern.

Example 1: Pathology reads the tumor is adenocarcinoma, acinar predominant (acinar 60%, solid predominant 20%, lepidic predominant 20%). Code the histology with the highest percentage: acinar adenocarcinoma 8551/3.

Example 2: Pathology reads the tumor is adenocarcinoma, solid predominant (with acinar, lepidic, and papillary subtypes). Code the predominant histology: solid adenocarcinoma 8230/3.

Example 3: Pathology reads the tumor is adenocarcinoma, lepidic prominent 80%, solid predominant 20% and the synoptic report states lepidic pattern 80%, solid pattern 20%. Code the histology with the higher pattern percentage: lepidic adenocarcinoma 8250/3.

Rule H8 Code a **combination** code when there are multiple histologies **AND**

- The combination is listed in **Table 2** in Equivalent Terms and Definitions, the ICD-O and all updates, **OR**
- You received a combination code from **Ask a SEER Registrar**.

Note: The rules are **hierarchical.** Use this rule only when previous rules do not **apply.**

Lung Histology Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule H9 Code adenocarcinoma with mixed subtypes **8255** for

- Multiple adenocarcinoma subtypes **OR**
- Any combination of histologies which are not listed in Table 2 in the Equivalent Terms and Definitions.

Note 1: Any combination of histologies listed in H7 with equal percentages is coded 8255.
Note 2: Adenocarcinoma with mixed subtypes 8255 does not apply to squamous cell carcinoma.

This is the end of instructions for Single Tumor

Code the histology using the rule that fits the case.

Lung Histology Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multiple Tumors Abstracted as a Single Primary

Note: Before coding histology, use the **Multiple Primary Rules** to determine that multiple tumors are a single primary.

Rule H10 Code **mucinous** adenocarcinoma (for lung only) when all tumors consist of:

- **8253/3** when
  - Behavior unknown/not documented (use staging form to determine behavior when available)
  - Invasive
- **8257/3** when
  - Microinvasive
  - Minimally invasive
- **8253/2** when
  - Preinvasive
  - In situ

Note 1: These are **new codes and terms** which will allow mucinous adenocarcinoma/carcinoma to be analyzed separately from colloid carcinoma.

Note 2: Changes take place over time. **Pathologists may not use** terms "minimally invasive" and "pre-invasive" **immediately**. **Code** the **pathology diagnosis.**

Rule H11 Code **non-mucinous** adenocarcinoma (for lung only) when all tumors consist of:

- **8256/3** when
  - Microinvasive
  - Minimally invasive
- **8250/2** when
  - Preinvasive
  - In situ

Note: These are new codes and terms.

Rule H12 Code the specific histology when the diagnosis for the tumor is **non-small cell lung carcinoma** (NSCLC) **consistent with** (or any other ambiguous term) **a specific carcinoma** (such as adenocarcinoma, squamous cell carcinoma, etc.) when:

- The histology is clinically confirmed by a physician (attending, pathologist, oncologist, pulmonologist, etc.)
- The patient is treated for the histology described by an ambiguous term
- The case is accessioned (added to your database) based on a single histology described by ambiguous terminology and no other histology information is available/documented

Note: If the case does not meet the criteria in the first two bullets, code non-small cell lung cancer (NSCLC) **8046**.

Example 1: Only one tumor is biopsied. The pathology diagnosis is NSCLC consistent with adenocarcinoma. The oncology consult says the patient has adenocarcinoma of the right lung. This is clinical confirmation of the diagnosis, code adenocarcinoma. The case meets the criteria in bullet 1.

Example 2: Only one tumor is biopsied. The pathology diagnosis is NSCLC consistent with squamous cell carcinoma. The treatment plan says the patient will receive the following treatment for squamous cell carcinoma. Treatment plan confirms squamous cell carcinoma; code squamous cell carcinoma. The case meets the criteria in bullet 2.

Example 3: Only one tumor is biopsied. Outpatient biopsy says probably squamous cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology squamous cell carcinoma. The case meets the criteria in bullet 3.

Rule H13 Code the histology when only **one** histology is present in **all** tumors.

Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in **Table 3** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Table 3,** use the **ICD-O** and all **updates**.

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

Rule H14 Code the **invasive** histology when all tumors have both invasive and in situ elements.

Note 1: All tumors may be **mixed** in situ and invasive **OR** one tumor may be in situ and the other invasive.

Note 2: Tumors may be **NOS** and a **subtype/variant**.

Note 3: When the NOS is invasive and the subtype/variant is situ, code the NOS (invasive).

Note 4: Multiple Primary Rules must be applied to be certain all tumors are a single primary.

Rule H15 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Mucinous adenocarcinoma and a subtype/variant of mucinous adenocarcinoma
- Non-small cell carcinoma **8046** and a subtype/variant of non-small cell carcinoma
- Sarcoma **8800** and a subtype/variant of sarcoma
- Neuroendocrine carcinoma (NEC) **8246** and a subtype/variant of NEC
- Neuroendocrine tumor (NET) **8240** and a subtype/variant of NET
- Squamous cell carcinoma **8070** and a subtype/variant of squamous cell carcinoma

Note 1: All tumors may be **mixed** histologies (NOS and a subtype/variant of that NOS) **OR** one tumor may be a **NOS** histology and the other tumor a **subtype/variant** of that NOS.

Note 2: See **Table 3** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

Rule H16 Code the appropriate **combination code** when all tumors have multiple histologies **AND**

- The combination is listed in **Table 2** in Equivalent Terms and Definitions, the ICD-O and all updates, **OR**
- You received a combination code from **Ask a SEER Registrar.**

Note: The rules are hierarchical. Use this rule **only** when previous rules do not apply.

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

Code the histology using the rule that fits the case.

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

Introduction

Note 1: This section includes the following **primary sites**: Peripheral nerves C470-C479; cerebral meninges C700; spinal meninges C701; meninges NOS C709; brain C710-C719; spinal cord C720; cauda equina C721; olfactory nerve C722; optic nerve C723; acoustic nerve C724; cranial nerve NOS C725; overlapping lesion of brain and central nervous system C728; nervous system NOS C729; pituitary gland C751; craniopharyngeal duct C752; pineal gland C753.

Note 2: Non-malignant intracranial and CNS tumors have a separate set of rules.

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

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

Note 4: There **must be** a histologic, cytologic, radiographic, or clinical **diagnosis** of a **malignant** neoplasm /3.

Note 5: Tumors from a number of primary sites metastasize to the brain. Do not use these rules for tumors described as metastases; report metastatic tumors using the rules for that primary site.

Note 6: **Pilocytic astrocytoma/juvenile pilocytic astrocytoma** is reportable in North America as a **malignant** neoplasm 9421/3.

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

Note 7: Tables and rules refer to ICD-O rather than ICD-O-3. The specific version is not specified to allow for updates. Use the currently approved version of ICD-O and all updates.

Note 8: For those sites/histologies which have recognized **biomarkers**, the biomarkers may aid in the identification of histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Note 9: See the Head and Neck Rules for coding paragangliomas.

Note 10: Use Malignant CNS Rules for histologies other than paraganglioma arising in C47_.

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Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Cerebrospinal fluid; CSF
- Dura; meninges
- Extradural; not within the meninges; within the cranium; within the skull but not within the cerebral meninges
- Extramedullary; outside medulla oblongata C700
- Infratentorial; below the tentorium cerebelli; either cerebellum or brainstem
- Intracranial; within the skull, within the cranium
- Intradural; between layers of the cerebral meninges; C700
- Intradural-extramedullary; within the spinal canal but outside of the nerves
- Intraspinal; occurring within the spinal column especially the vertebral canal; spinal nerve roots
- Site; topography
- Supratentorial; above the tentorium cerebelli; cerebrum
  - Cerebrum contains frontal lobe, parietal lobe, occipital lobe, and temporal lobe
- Tentorium cerebelli; cerebellar tentorium
- Tumor; mass; lesion; neoplasm when
  - These terms are used ONLY to determine multiple primaries
  - Do not use these terms for casefinding or determining reportability
- Type; subtype; variant
- WHO Grade 3 and WHO Grade 4; malignant; invasive; /3

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Terms that are NOT Equivalent or Equal

This is a term that is **not equivalent**. There are no casefinding implications.

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

Changes from 2007 MPH Rules

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

1. 2016 CNS WHO presents major restructuring of the diffuse gliomas, medulloblastomas and other embryonal tumors, and incorporates new entities that are defined by both histology and molecular features, including glioblastoma, IDH-wildtype and glioblastoma, IDH-mutant; diffuse midline glioma, H3 K27M-mutant, RELA fusion-positive ependymoma, medulloblastoma, WNT-activated and medulloblastoma, SHH-activated, and embryonal tumor with multilayered rosettes, C19MC-altered. The 2016 edition has added newly recognized neoplasms and has referred to some entities, variants and patterns as "not recommended" (previously called obsolete).
   A. It has been determined that these "not recommended" terms no longer have diagnostic and/or biological relevance. For example, gliomatosis cerebri is a term which is no longer recommended. Gliomatosis cerebri is now termed a "growth pattern" rather than a histologic type.
   B. Terms which are not recommended are not included in the tables. When one of these terms are used, refer to the ICD-O and all updates for the correct histology code. For example, glioma NOS is an umbrella term for all gliomas and astrocytomas. Glioma NOS is not recommended because diagnostic methodology is able to determine a more specific diagnosis.
2. **Rule change:** The 2007 rules said a glioblastoma multiforme (GBM) following an astrocytic or glial tumor was a single primary (recurrence). In the 2018 Solid Tumor Rules, GBM subsequent to an astrocytic or glial tumor is a multiple primary. GBM is now being collected as a new primary so it is possible to analyze the frequency with which these tumors recur in a more aggressive form (GBM).

3. **Clarifications:**
   A. The following meningiomas are reportable: intraosseous, cavernous sinus and sphenoid wing.
   B. Multiple cerebral meningiomas are a single primary.
   C. Multiple brain tumors (same histology) are a single primary.
   D. Laterality is not used to determine multiple primaries.
   E. Timing is not used to determine multiple primaries.
   F. The brain (C710-C719) is a single primary site.
   G. Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are genetic syndromes and not reportable neoplasms. People with this genetic syndrome do have a high risk of developing:
      i. Non-reportable non-malignant tumors occurring in skin and sites other than CNS AND
      ii. Reportable malignant tumors
4. The 2016 World Health Organization (WHO) Classification of Tumors of the Central Nervous System (CNS) contains newly recognized histologies and subtypes/variants. New codes/terms are identified by asterisks (*) in **Table 3** in the Terms and Definitions.

New for 2023

1. Beginning with cases diagnosed 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma will **no longer be reported as malignant (/3)**. These neoplasms will continue to be reportable with a behavior of /1. Historically, pilocytic astrocytomas were coded as 9421/3 from 1976-2000. Beginning in 2001 with the release of ICD-O-3, WHO changed the behavior to /1, however, the standard setters opted to continue collecting these cases using malignant /3 behavior. This practice will continue through 12/31/2022.
2. WHO 5th Ed CNS Tumors has assigned a new histology to 9421/3. Beginning with cases diagnosed 1/1/2023, for surveillance purposes, code 9421/3 will be valid for the following histology ***only:***
   A. High Grade astrocytoma with piloid features (HGAP)

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Reportability Criteria

CNS neoplasms must meet all **three of the conditions below** to be **reported as malignant /3**:

1. The **behavior** must be malignant /3:
   A. Pathology designates the behavior as malignant/invasive, /3 **OR**
   B. The tumor is a WHO Grade 3 or 4 (See **Section 1: Table 1**)

   Note 1: WHO Grade 2 tumors may be non-malignant or malignant.
   Note 2: **Always** code the behavior as designated by the **pathologist.**
   Note 3: **Never report** a **malignant** /3 behavior code for a meningioma **based on tumor extension** to brain, skin of scalp, or other regional organs/tissue. Non-malignant CNS tumors can extend to the regional tissue and bone.

2. The **primary site** must be reportable (See **Section 2: Table 2**) **AND**
3. The **histology** must be reportable (See **Section 2: Table 3**)

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Directory of Sections and Tables

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

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Section 1: Behavior Code

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

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

Priority Order for Using Documentation to Assign Behavior

1. Pathology: Tissue from **resection**
   A. Use the pathologist's **description** of **malignant/invasive** behavior
   B. Cases are reportable as malignant when pathology states a WHO Grade 3 or 4
      i. WHO Grade 2 may be either non-malignant or malignant. Use the pathologist's description of behavior (priority #1).
   C. **Never change behavior** described by pathologist
2. Pathology: Tissue from **biopsy**
3. Cytology (usually cerebrospinal fluid)
4. Physician's documentation (no pathology report) in the following priority order:
   A. Tumor Board
   B. Documentation of original pathologic diagnosis and behavior
      Example: Pathology report is not in the medical record. Oncology consult states biopsy was done in a different facility and cites the **original pathology** diagnosis including the **behavior**.
   C. **Documentation** of behavior, no mention of original diagnosis
      Example: Pathology report is not in the medical record. **Physician documents** the **behavior** as malignant, or WHO Grade 3 or 4, but **does not cite/mention original** pathology report as source of behavior classification.
5. **Scans**, in the following **priority order**:
   A. MRI
   B. CT
   C. PET
   D. Angiogram
6. When instructions **1-5 do not apply**, use **Table 1** to determine behavior.

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Table 1: WHO Grades for Select CNS Neoplasms

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

WHO Grade Definitions

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

WHO Grade II CNS Tumors: Non-malignant and Malignant

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

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

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

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

Table Instructions

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

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

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

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

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Histology	WHO Grade
Malignant peripheral nerve sheath tumor (MPNST) ***Note:*** Tissue/pathology reports or CAP protocol/summary will specify whether this is WHO Grade 2, 3, or 4	2, 3, or 4
Medulloblastoma (including all subtypes)	4
Medulloepithelioma	4
Meningioma	1
Myxopapillary ependymoma	1
Neurofibroma	1
Oligodendroglioma IDH-mutant and 1p/19q deleted	2
Papillary glioneuronal tumor	1
Papillary tumor of the pineal region ***Note:*** Tissue/pathology reports or CAP protocol/summary will specify whether it is WHO Grade 2 or 3	2 or 3
Perineuroma	1
Pilocytic astrocytoma ***Note:*** Collected as malignant /3 in North America	1
Pineal parenchymal tumor of intermediate differentiation ***Note****:* Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 2 or 3	2 or 3
Pineoblastoma	4
Pineocytoma	1
Pituicytoma	1
Pleomorphic xanthroastrocytoma	2
Rosette-forming glioneuronal tumor	1
Schwannoma	1

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

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Section 2: Reportable Primary Sites and Histologies

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

Note 1: **Peripheral nerves** are included in the Malignant CNS and Peripheral Nerve rules because:

- **All malignant** tumors are reportable, including peripheral nerve tumors **AND**
- The Malignant CNS and Peripheral Nerve **rules** contain the **correct histologies** and coding **rules** for tumors of **peripheral nerves and meninges/dura.**

Note 2: **Peripheral nerves** are:

- **Extracranial**/outside the cranium **OR**
- **Extradural**/outside the spinal dura

Note 3: The following malignant **meningiomas** are reportable:

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

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Priorities for Coding Primary Site

Note 1: **Always** check the **operative report(s)** which will have **information** on whether the surgery or biopsy was **intracranial** (inside the cranium/skull) or **intraspinal** (within the dura/meninges covering the spinal cord).
Note 2: Code **the specific primary site**. Use an **NOS** site code **only** when a specific site is not known.

Use the list in **hierarchical order**:

1. **Resection**
   A. Operative report(s)
   B. Pathology report(s)
2. **Biopsy**
   A. Operative report(s)
   B. Pathology report(s)
3. Resection and/or biopsy performed, but operative report(s) and pathology are not available (minimal information)
   A. Tumor Board
   B. Code from physician's documentation of **original diagnosis** from operative or pathology report OR
   C. Physician's **documentation of primary site** in the medical record
   Example: The patient had a **biopsy** done at **another facility**. The operative and pathology reports are not in the medical record. The **attending documents** that the patient had a biopsy of a **cerebral meningioma** at a different facility. Code the primary site cerebral meninges C700.
4. For cases diagnosed by imaging (**no pathology/resection or biopsy**) use information from scans in the following priority order:
   A. MRI
   B. CT
   C. PET
   D. Angiogram
5. See **Table 2**: **Reportable Primary Sites** to confirm the primary site is reportable.
6. When the primary site is cranial nerve **OR** peripheral nerve, see **Table 4**: **Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves** to determine whether the portion of the nerve is cranial or peripheral (different site codes).

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Reportable Primary Site Groups

The three major groups of reportable sites are:

1. **Intracranial** (within the skull/cranium) **AND**
2. **Spinal sites** (spinal meninges and sites within the spinal meninges)
3. **Peripheral nerves** (extracranial or extraspinal nerves)**.**

Terms and sites for these broad categories are:

Reportable Primary Sites and their ICD-O Codes

1. Intracranial (intra means inside or within; intracranial is inside the skull).
   A. **Cerebral/cranial dura/meninges** C700. The cerebral meninges has three layers:
      i. **Dura** mater is the **superficial** layer of meninges
         - Tightly adherent to skull
         - Contains folds and **sinuses**
         - Contacts **endosteum** which lines the bones of the skull
      ii. **Arachnoid** mater forms the middle of the three layers of meninges
      iii. **Pia** mater is the delicate vascular fibrous membrane which is adherent to the brain.
   B. **Brain** C710-C719
   C. **Cranial nerves** C722-C729. See **Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves**
   D. **Intracranial glands C751-C753**
      i. Craniopharyngeal duct C752
      ii. Pineal gland C753
      iii. Pituitary gland C751

Continued on next page

2. **Spinal** sites: includes the **spinal meninges** and **all** structures **within** the meninges (Intradural, within/in the spinal meninges).
   A. **Spinal cord C720**
   B. **Spinal meninges C701** covers/encloses the spinal nerve roots and the spinal cord.
   C. Spinal nerve roots:
      i. **Cervical** nerve, 8 pair, occipital nerve. Code to peripheral nerve of head, face, and neck C470
      ii. **Coccygeal** nerve, 1 pair. Code to cauda equina C721
      iii. **Lumbar** nerve, 5 pair. Code to cauda equina C721
      iv. **Sacral** nerve, 5 pair. Code to cauda equina C721
      v. **Thoracic** nerve, 12 pair. Code to peripheral nerves of thorax C473
3. **Peripheral nerves**
      i. **Cervical** nerve, 8 pair, occipital nerve. Code to peripheral nerve of head, face, and neck C470
      ii. **Thoracic** nerve, 12 pair. Code to peripheral nerves of thorax C473
      iii. **Lumbar** nerve, 5 pair. Code to cauda equina C721
      iv. **Sacral** nerve, 5 pair. Code to cauda equina C721
      v. **Coccygeal** nerve, 1 pair. Code to cauda equina C721

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Table 2: Reportable Primary Sites

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

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

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Site Group	Reportable Subsite Terms and Code
Ill-Defined Sites Central Nervous System	Nervous system NOS **C729** Overlapping lesion of brain and central nervous system **C728**
Intracranial Duct and Glands	Craniopharyngeal duct **C752** Pineal gland **C753** Pituitary gland **C751**
Meninges	Cerebral meninges **C700** Meninges NOS **C709** Spinal meninges **C701**
Peripheral Nerve and Autonomic Nervous System	Abdomen **C474** Autonomic nervous system NOS **C479** Head, face and neck **C470** Lower limb and hip **C472** Nerves of pelvis **C475** Overlapping lesion of peripheral nerves and autonomic nervous system **C478** Thorax **C473** Trunk NOS **C476** Upper limbs and shoulder **C471** Spinal Nerve NOS **C479**
Spinal Sites	Cauda equina/ **C721** Conus medullaris/filum terminale **C720** Meninges NOS **C709** Spinal cord **C720** Spinal meninges **C701**

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

Table 3 lists the more common histologies for CNS and Peripheral Nerves. For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database.**

Note: Behavior codes are not listed because all histologies are **malignant /3.**

Column 1 contains specific and NOS histology terms and codes.

- Specific histology terms do not have subtypes/variants
- **NOS** histology terms **do** have **subtypes/variants**

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

Note: All tumors are malignant/invasive /3.

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

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

Table begins on next page

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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Anaplastic ganglioglioma **9505**		
Astroblastoma **9430**	Astroblastoma, MN1-altered	
Astrocytoma NOS **9400**	Astrocytoma, IDH-mutant, grade 2 Diffuse astrocytoma IDH-mutant Diffuse astrocytoma IDH-wildtype Diffuse astrocytoma NOS	Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401** Astrocytoma, IDH-mutant, grade 3 **9401** Astrocytoma, IDH-mutant, grade 4 **9445** Gemistocytic astrocytoma IDH-mutant **9411** Pleomorphic xanthroastrocytoma /anaplastic pleomorphic xanthroastrocytoma **9424**
Cauda equina neuroendocrine tumor **8693/3** ***Note***: This neoplasm is coded with /3 behavior even though it is a WHO Grade 1.		
Choriocarcinoma **9100**		
Choroid plexus carcinoma **9390**		
CNS embryonal tumor with rhabdoid features **9508**	Atypical teratoid/rhabdoid tumor Embryonal tumor with rhabdoid features	
CNS ganglioneuroblastoma **9490**		CNS embryonal tumor, NEC/NOS **9473**
CNS neuroblastoma **9500**	CAN neuroblastoma, FOXR2-activated CNS Tumor with BCOR internal tandem duplication	

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Diffuse leptomeningeal glioneuronal tumor **9509*** ***Note 1***: Cases diagnosed prior to 1/1/2023 are coded 9509/1. See the non-malignant CNS rules. ***Note 2:*** Cases diagnosed 1/1/2023 forward are coded 9509/3.	DLGNT	
Diffuse midline glioma H3 K27M mutant **9385***	Diffuse intrinsic pontine glioma Diffuse hemispheric glioma, H3 G34-mutant Diffuse pediatric-type high grade glioma, H3-wildtype and IDH-wildtype DIPG Infant-type hemispheric glioma	
Embryonal carcinoma **9070**		Yolk sac tumor **9071**
Embryonal tumor with multilayered rosettes C19MC-altered **9478***	Embryonal tumor with multilayered rosettes, NOS ETMR	

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Ependymoma **9391** ***Note:*** The following terms are synonyms of ependymoma, RELA fusion-positive 9396, and are NOT subtypes/variants of it. They are all coded 9396. • Posterior fossa group A (PFA) ependymoma • Posterior fossa group B (PFB) ependymoma • Spinal ependymoma, MYCN-amplified • Supratentorial ependymoma, YAP1 fusion-positive • Supratentorial ependymoma, ZFTA fusion-positive	Clear cell ependymoma Posterior fossa ependymoma, NOS Spinal ependymoma, NOS Supratentorial ependymoma, NOS Tanycytic ependymoma	Anaplastic ependymoma **9392** Ependymoma, RELA fusion-positive **9396*** Posterior fossa group A (PFA) ependymoma Posterior fossa group B (PFB) ependymoma Spinal ependymoma, MYCN-amplified Supratentorial ependymoma, YAP1 fusion-positive Supratentorial ependymoma, ZFTA fusion-positive Papillary ependymoma **9393**
Epithelioid hemangioendothelioma **9133**		
Germinoma **9064**		
Glioblastoma NOS **9440**	Glioblastoma multiforme GBM Glioblastoma, IDH wild-type Epithelioid glioblastoma	Giant cell glioblastoma **9441** Glioblastoma IDH-mutant **9445*** Gliosarcoma **9442**

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Glioma, NOS **9380** ***Note***: Glioma, NOS is considered an umbrella term. Additional testing should be performed to identify mutations and biomarkers that would provide a definitive type. A diagnosis of glioma, NOS is not recommended and may be used only when additional test were inconclusive. **IMPORTANT**: See M rules to determine multiple primaries.		
High-grade astrocytoma with piloid features **9421/3** ***Note***: This term is reportable for cases diagnosed 1/1/2023 forward.	HGAP	
Immature teratoma **9080**		Mixed germ cell tumor **9085** Teratoma with malignant transformation **9084**
Malignant meningioma **9530**	Anaplastic meningioma	Papillary/rhabdoid meningioma **9538**
Malignant peripheral nerve sheath tumor **9540**	Malignant melanotic nerve sheath tumor Malignant perineurioma MPNST MPNST with perineural differentiation	Epithelioid malignant peripheral nerve sheath tumor **9542**

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Medulloblastoma NOS **9470**	Classic medulloblastoma Medulloblastoma, histologically defined	Anaplastic/large cell medulloblastoma **9474** Medulloblastoma described as one of the following **9471** Desmoplastic SHH-activated and TP53-wildtype With extensive nodularity Nodular Medulloblastoma non-WNT/non-SHH; medulloblastoma group 3 or group 4 **9477*** Medulloblastoma SHH-activated and TP53-mutant **9476*** Medulloblastoma WNT-activated **9475***
Medulloepithelioma **9501**		
Meningeal melanoma **8720**		Meningeal melanomatosis **8728**
Neuroepithelial tumor, malignant **8000/3** ***Note***: Neuroepithelial tumor is a rare tumor specific to children. These neoplasms have numerous subtypes which are not easily identified so a specific type may not be identified on the pathology report. WHO has not proposed an ICD-O code for this entity. The current option is to assign code 8000. Because these tumors are different, they are on a separate row.		

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Oligoastrocytoma NOS **9382**	Anaplastic oligoastrocytoma NOS	
Oligodendroglioma NOS **9450** ***Note:*** Oligodendroglioma NOS is used when molecular markers cannot fully be determined	Oligodendroglioma 1p/19q-codeleted Oligodendroglioma IDH-mutant Oligodendroglioma IDH-mutant and 1p/19q-codeleted, grade 2	Anaplastic oligodendroglioma NOS **9451** IDH-mutant 1p/19q-codeleted IDH-mutant and 1p/19q-codeleted Oligodendroglioma, IDH-mutant and 1p/19q-codeleted, grade 3
Peripheral primitive neuroectodermal tumor **9364**	Ewing sarcoma pPNET	
Pilocytic astrocytoma **9421** ***Note 1:*** ICD-O-3 lists a behavior code of /1. It is collected as /3 in North America for cases diagnosed prior to 1/1/2023. ***Note 2***: Beginning with cases diagnosed 1/1/2023 forward, pilocytic astrocytoma will no longer be reported with /3 behavior. Pilocytic astrocytoma and the related terms listed below are to be reported as a /1 • Diffuse astrocytoma, MTB- or MYBL1-alterd • Diffuse low-grade glioma, MAPK pathway- altered+		Pilomyxoid astrocytoma **9425**
Pineal parenchymal tumor of intermediate differentiation **9362**	Pineoblastoma	Papillary tumor of the pineal region **9395**
Pituitary adenoma/pituitary neuroendocrine tumor **8272/3**	PitNET	

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

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

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

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

Note 1: Neoplasms arising in a **cranial** or **spinal** nerve are coded to the specific nerve in which it arises.
Note 2: Neoplasms, commonly meningiomas, arising in the **dura/meninges** of an **intracranial** nerve (cranial nerve within the skull) are coded to **cerebral meninges** C700.
Note 3: Neoplasms, commonly meningiomas, arising in the **dura/meninges** of the **spinal nerve roots** are coded to the ICD-O site code **spinal meninges** C701.
Note 4: It is important to **check** the **operative report** to determine whether the surgery is **intracranial** or **intradural.**

Use **Table 4** to **determine** whether a malignant cranial nerve tumor should be coded to a cranial nerve or to a peripheral nerve. The anatomic location of the tumor determines whether the primary site is a peripheral or a cranial nerve.

Column 1: The **proper name** for the cranial nerve (CN) and the **cranial nerve number**
Column 2: The point at which the nerve exits the cranium
Column 3: Portions of the nerve coded to cranial nerve
Column 4: Portions of nerve coded to peripheral nerve

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

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Trochlear CN 4	Superior orbital fissure	Arises from the **dorsal brain stem**, loops around the brainstem and passes anteriorly within the **subarachnoid space**. It travels between the **superior cerebellar and posterior cerebral arteries** and through the **dura,** enters **cavernous sinus C725**	Enters the orbital fissure **C470**
Trigeminal CN 5 ***Note:*** Trigeminal is derived from Latin trigeminus which means **born in threes** (tri) and born **at the same time** (germinal). As the name implies, the nerve **separates** into **three branches**; ophthalmic, maxillary, and mandibular	The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina. Superior orbital fissure. The foramen rotundum The foramen ovale.	CN5 originates in the **pons**. Upon leaving the pons it enters a **small fossa** posterior and inferolateral to the cavernous sinus called **Meckel's (trigeminal) cave C725** .	• **Ophthalmic nerve branch** crosses the **pterygopalatine fossa**, inclines laterally on the back of the **maxilla**, and enters the **orbit** through the inferior orbital fissure where it is called the **infraorbital** nerve. It ends beneath the **quadatus labii superius**, and divides into multiple branches that spread to the side of the **nose**, the lower **eyelid**, and the upper **lip C470** • **Maxillary** nerve leaves foramen rotundum and traverses the **infraorbital groove** and **canal** in the floor of the **orbit**, and appears on the face at the infraorbital foramen **C470** • **Mandibular** nerve leaves via the foremen ovale travels along the **mandibular** groove **C470**

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

Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Abducent CN 6	Cranial meninges	Exits brainstem at junction of **pons** and the **medulla**, enters the **subarachnoid** space and runs upward between the pons and the **clivus** entering the **cavernous sinus C725**	**Dorello's canal** and travels to the tip of the **temporal bone**. Enters **orbit C470**
Facial CN 7	Internal acoustic meatus	**CN7** originates in the **pons,** along the posterior cranial fossa (**posterior cranial fossa** (the posterior cranial fossa is part of the intracranial cavity.) **C725**	Enters the temple through the internal auditory meatus and runs through the facial canal. **C470**
Acoustic or vestibulocochlear CN 8	Internal acoustic meatus	Originates in the **brain stem** (**medulla oblongata**) **between** the base of the brain (**pons**) and the **spinal cord C724** Both the **vestibular** branch and the **cochlear** branch are located in the **inner ear**	
Glossopharyngeal CN 9	Jugular foramin[a]	**Originates** in the anterior portion of the **medulla oblongata C725**	Jugular foramen Between the i**nternal jugular** vein and **internal carotid** artery Lies on the **stylopharyngeus** and middle **pharyngeal** constrictor muscle Passes under the **hypoglossus** muscle Palatine tonsil Extends to mucous **glands** of the **mouth**, and **base** of the **tongue C470**

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

Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Vagus CN 10	Jugular foramen	The vagus nerve originates from the **medulla** of the **brainstem**. **C725**	CN10 descends within the **carotid sheath** medial to the internal jugular vein at the root of the **neck C470**. The right vagus crosses in front of the subclavian artery and travels into the **fat behind** the blood **vessels**, reaching the **thorax**. It then inclines **behind** the **hilum** of the **right lung** and moves toward the esophagus. The nerve splits into the **right** and **left** vagus at the **esophageal plexus C473** Forms the anterior and posterior **gastric** nerves **C475**
Accessory CN 11	Jugular foramen	The spinal accessory nerve **originates** in the neurons of the upper spinal cord, specifically **C1-C5/C6** spinal **nerve roots**. The nerve enters the foramen magnum or lateral aspect of the **medulla oblongata.** The fibers of the spinal accessory nerve coalesce to **form spinal rootlets, roots**, and finally the **spinal accessory nerv**e itself C**725**	The nerve **exits** the skull through the jugular foramen. It then runs along the internal **carotid** artery within the **neck C470** Reaches the sternocleidomastoid muscle and the trapezius **C476**
Hypoglossal CN 12	Hypoglossal canal	**CN12** starts in the **hypoglossal** nucleus of the **brainstem**, **C725**	**CN12** exits the hypoglossal canal, traveling **between** the **carotid** artery and **jugular** vein, ending under the **tongue C470**

Section 3: Additional Information to Complete the Abstract

Conflicting Information on Pathology Report(s)

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

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

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

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

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

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

Table 5: Paired Sites

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

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

Note 1: Midline tumors are common for glioblastoma multiform and meningiomas.
Note 2: SEER allows laterality to be coded for sites other than those in the table.

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

Table 6: Non-malignant CNS Tumors with the Potential of Transforming to Malignant Behavior

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

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

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

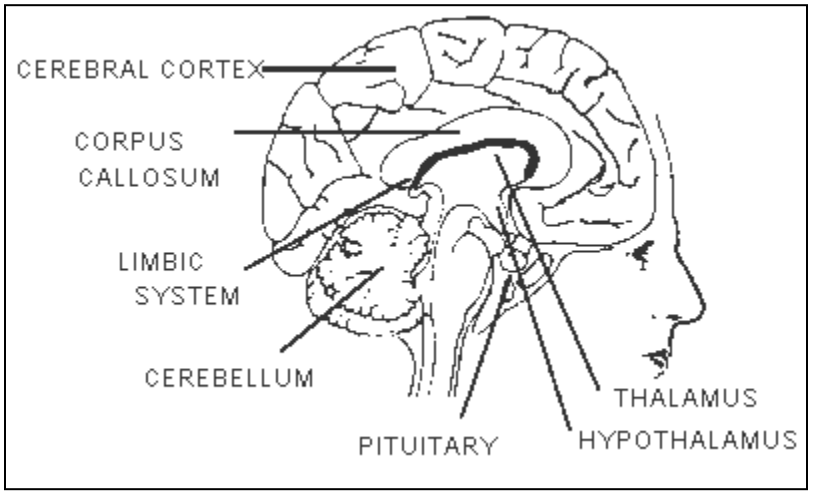
Use **Table 6** when directed to by the Multiple Primary Rules.

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

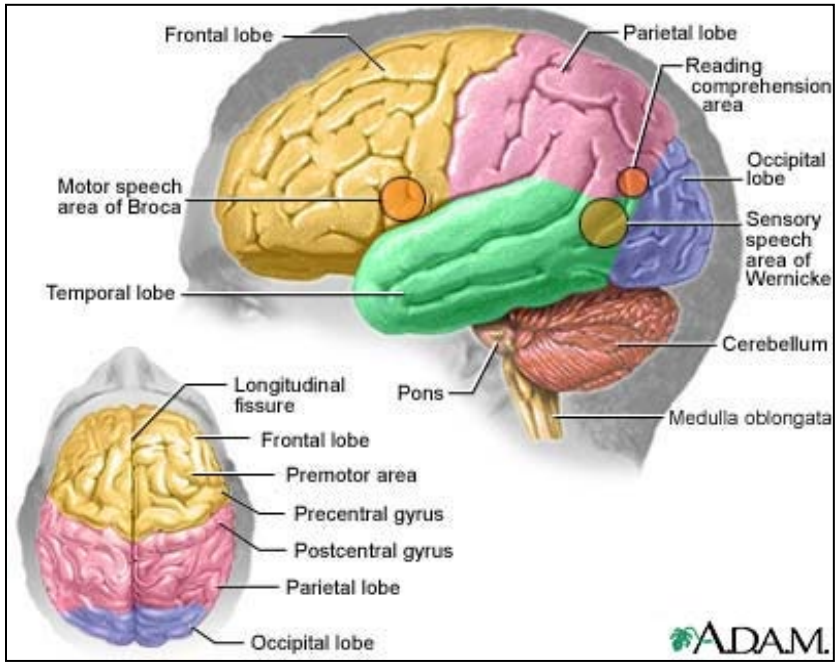
Original Histology and Code	Transformed Histology and Code
Chondroma **9220/0**	Chondrosarcoma **9220/3**
Ganglioglioma **9505/1**	Anaplastic ganglioglioma **9505/3**
Hemangioma **9120/0**	Angiosarcoma **9120/3**
Hemangiopericytoma **9150/1**	Anaplastic hemangiopericytoma **9150/3**
Leiomyoma **8890/0**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

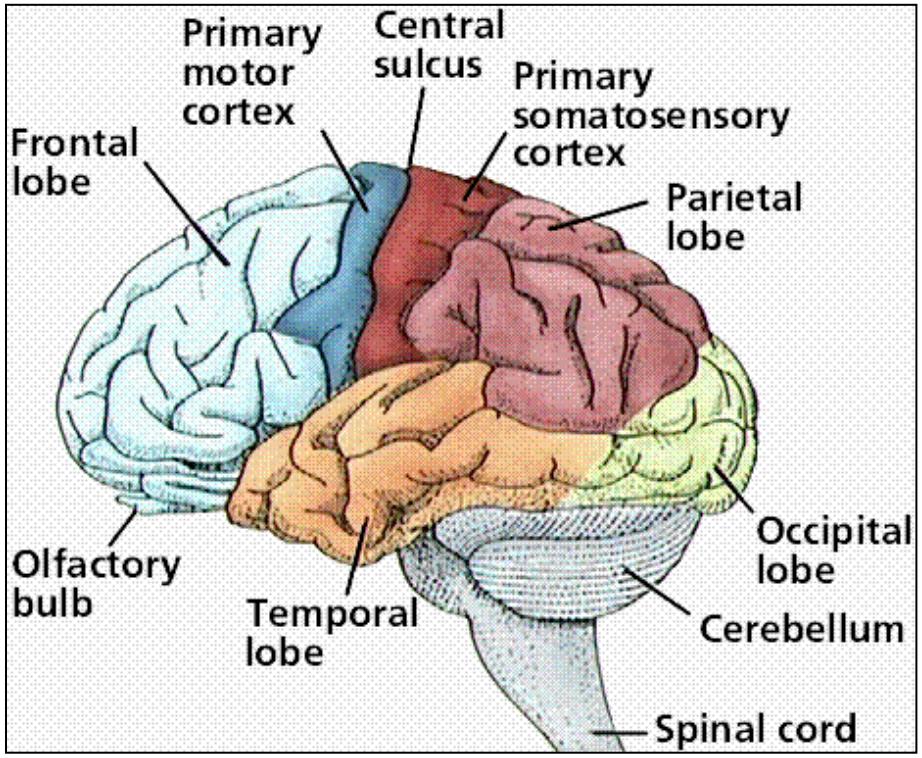
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Illustrations

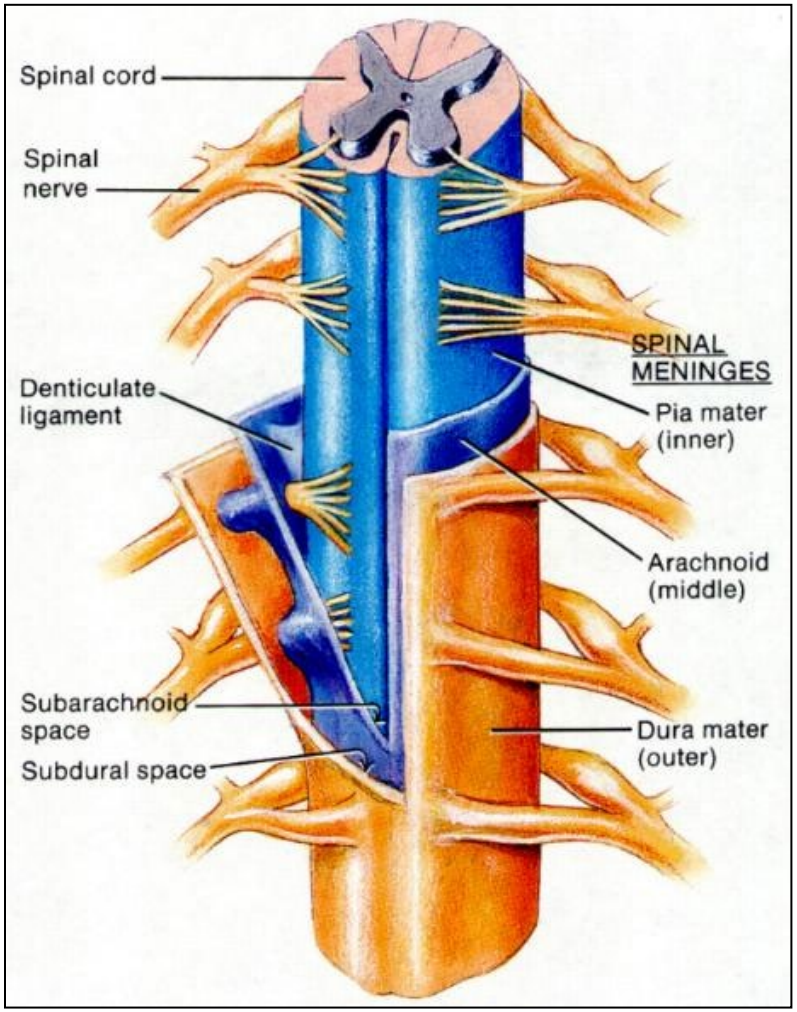


Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

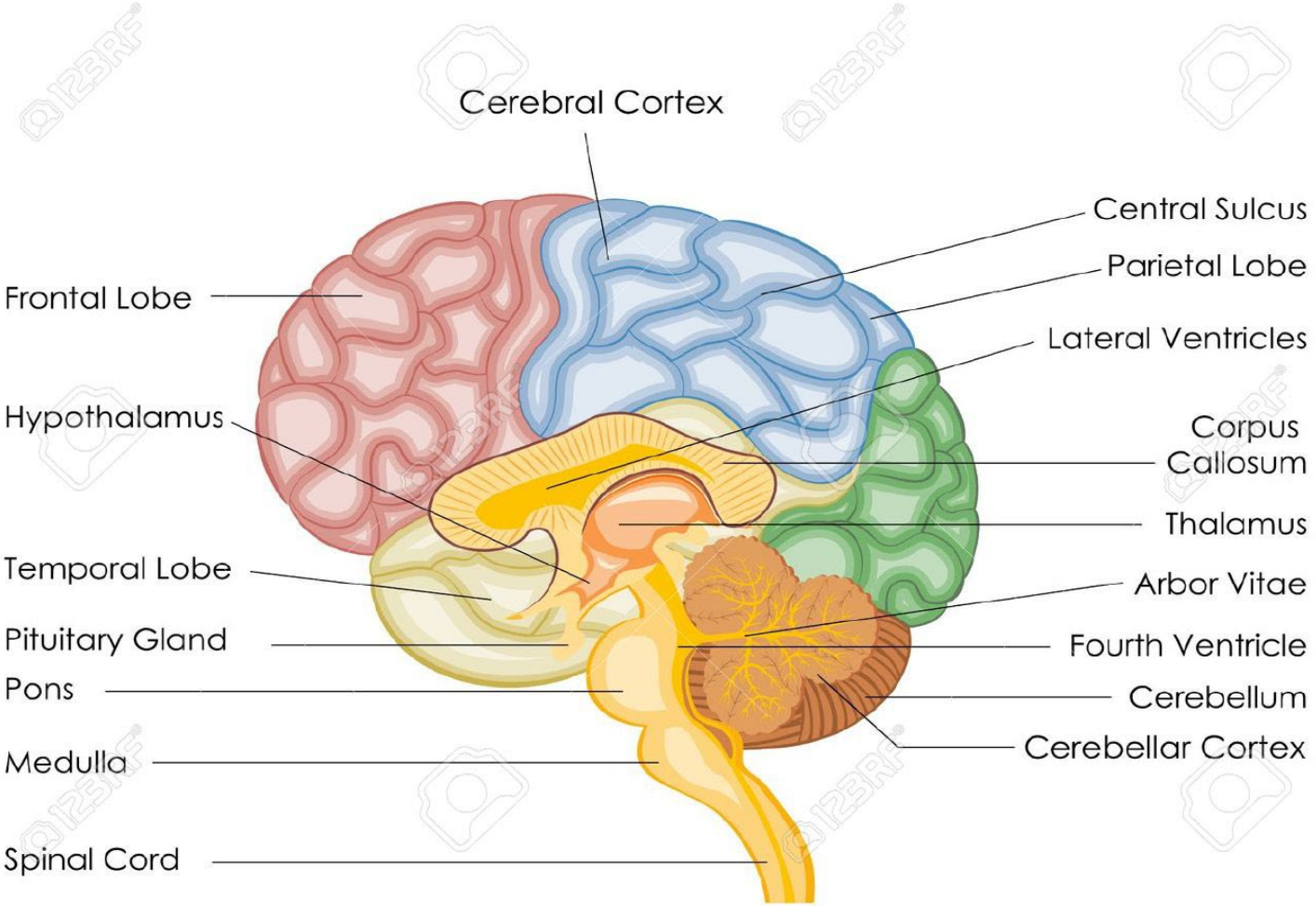




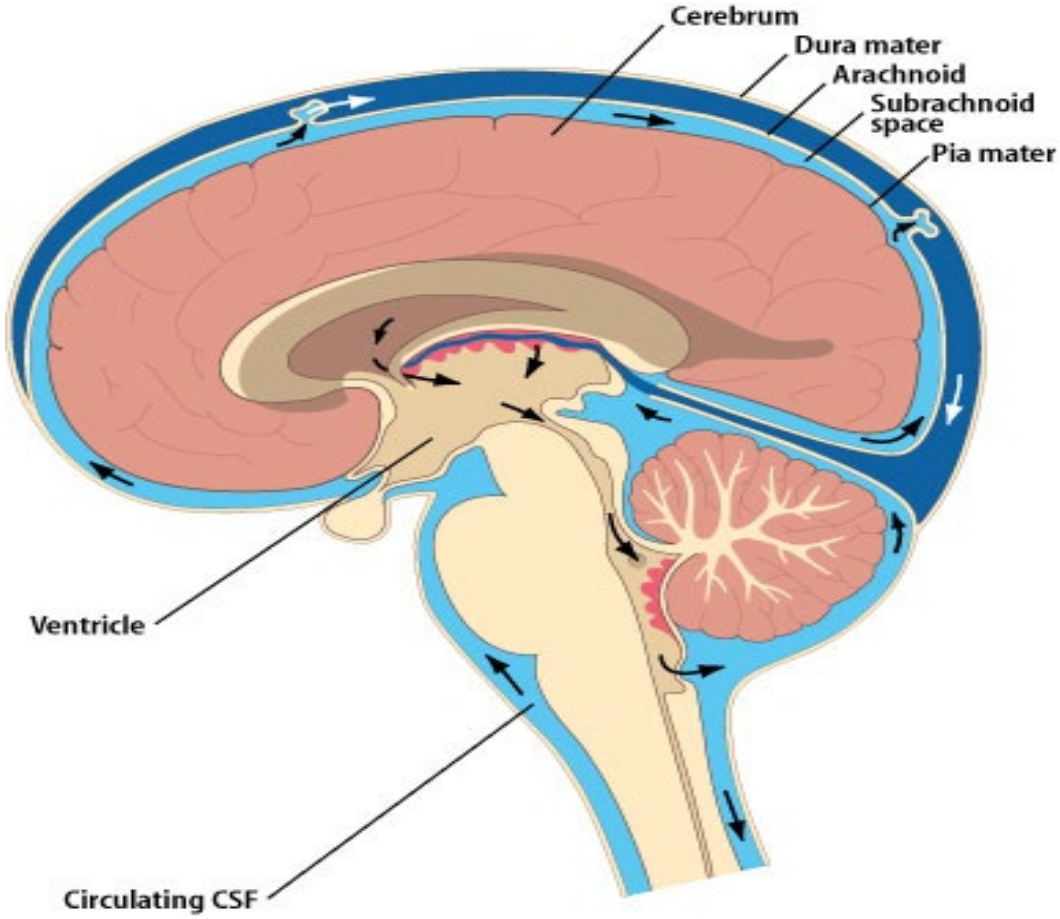
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



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



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



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

Note 1: Non-malignant intracranial and CNS tumors have a separate set of rules.
Note 2: Laterality **is not** used to determine multiple primaries for malignant CNS tumors.
Note 3: Timing **is not** used to determine multiple primaries for malignant CNS tumors.
Note 4: Separate GBM following an astrocytic or glial tumor is a multiple primary.
Note 5: These rules are **NOT** used for tumor(s) described as metastases.
Note 6: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown If Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor **or multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted
Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
- Outpatient biopsy with no follow-up information available
- Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors.

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

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

Single Tumor

IMPORTANT: The **major difference** between **M5** and **M6** is:
M5: No resection as first course of treatment **AND** when the same tumor is subsequently **resected**, **pathology proves malignant** behavior
M6: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo**) is **malignant**

Rule M2 Abstract a **single primary**[i] when there is a **single tumor**.

Note 1: A single tumor is always a single primary.
Note 2: The tumor **may overlap onto** or **extend** into **adjacent**/contiguous site or subsites.
Note 3: The tumor may have two or more histologic components.
Note 4: A glioblastoma multiforme 9440 (GBM) diagnosed in residual tumor, regardless of timing, is not a new primary

Rule M3 Abstract a **single primary**[i] when a neoplasm is **originally diagnosed** as an **oligodendroglioma** and subsequently **recurs** in residual tumor tissue with **different features** such as a densely cellular tumor with pseudo palisading necrosis.

Note 1: The pathology may state that the recurrence "looks like" or "has the appearance of" a glioblastoma multiforme (GBM). This is not a true GBM.
Note 2: Record as a recurrence for those registrars who collect recurrence data.

Rule M4 Abstract a **single primary**[i] when a neoplasm is originally diagnosed as **Glioma, NOS** and subsequently recurs in residual tumor with a more specific histology.

Note 1: Glioma, NOS is considered an umbrella term. Additional testing should be performed to identify mutations and biomarkers that would provide a definitive histology type. A diagnosis of glioma, NOS is not recommended and may be used only when additional tests were inconclusive.
Note 2: If a specific histology is diagnosed in residual tumor or additional testing provides a definitive histology, edit the original abstract as follows:

- Do not change the date of diagnosis
- For cases that have been abstracted, update the ICD-O code based on the new findings
- Report all data changes for cases which have been submitted to the central registry

Note 3: There is no time requirement.

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

Rule M5 Abstract a **single primary**[i] (the malignant) when a single tumor meets the following two criteria:

1. The original diagnosis was non-malignant /0 or /1 **AND**
   - First course treatment was active surveillance (no tumor resection). Diagnosis was:
     - Clinical
     - Radiographic
     - Stereotactic biopsy
2. Subsequent resection pathology is malignant /3

Note 1: This rule **clarifies** that a single tumor is **always** a single primary and the malignant behavior is reported.

Note 2: The **resection pathology** is **more accurate** than clinical, radiographic or stereotactic biopsy information. While stereotactic **biopsy provides** a **pathologic** specimen, it is small and **may not** include the **malignant** portion of tumor.

Note 3: There is **no time requirement** from initial diagnosis to resection.

Note 4: Edit the original abstract as follows:

- Do **not** change **date of diagnosis**.
- For cases which have been abstracted, **change behavior** code on original abstract from /0 or /1 to /3.
- **Report** all data changes for cases which have been submitted to the central registry.
- See the **COC** and **SEER manuals** for **instructions** on coding **other data items** such as Accession Year, Treatment and Sequence Number.

Note 5: The physician may have staged the non-malignant tumor at the time of original diagnosis (benign). The physician will also stage the malignant tumor. Staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology.

Example 1: A patient is diagnosed by **MRI** with **ganglioglioma 9505/1** in July 2018. After months of active surveillance (watchful waiting), the **patient** becomes **symptomatic**. A **resection** is done in April 2019; the **resection** pathology is **anaplastic ganglioglioma 9505/3**. **Change behavior code** on the **original abstrac**t from /1 to /3. Do not **change date of diagnosis.**

Example 2: A November 15, 2017 **MRI** and **subsequent** stereotactic **biopsy** of lateral ventricle of brain show a **mature teratoma 9080/1**. The patient becomes symptomatic in 2018 and the neoplasm is resected on October 31, 2018. The **resection pathology** diagnoses **immature teratoma 9080/3. Change behavior code** on the **original abstract. Do not change date of diagnosis.**

This is the end of instructions for Single Tumor.

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

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

Multiple Tumors

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

- In the CNS (see **Table 2**) AND in a site other than the CNS
  Example: Patient has a malignant nerve sheath tumor of the right arm C471 and non-metastatic adenocarcinoma of the lung. The lung is not a CNS site. Abstract two primaries.
- In different CNS sites (see Rule M9)

IMPORTANT: The **major difference** between **M5** and **M6** is:
M5: No resection as first course of treatment **AND** when the same tumor is subsequently **resected**, **pathology proves malignant** behavior
M6: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo)** is **malignant**

Rule M6 Abstract **multiple primaries**[ii] when there are multiple CNS tumors, one of which is malignant /3 and the other is non-malignant /0 or /1.

- Original non-malignant tumor followed by malignant tumor
  - Patient had a resection of the non-malignant tumor (not the same tumor) **OR**
  - It is unknown/not documented if the patient had a resection
- Simultaneous non-malignant and malignant tumors
  - Abstract both the malignant and the non-malignant tumors

Note 1: The rules are hierarchical. Only use when previous rules do not apply.
Note 2: See **Table 2** in the Equivalent Terms and Definitions for a listing of CNS sites.
Note 3: A non-malignant CNS tumor and a malignant CNS tumor are **always multiple** primaries (timing and primary sites are irrelevant). Prepare two abstracts; one for the non-malignant and another for the malignant tumor.

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

Rule M7 Abstract **multiple primaries**[ii] when a patient has a glial tumor and is subsequently diagnosed with a **glioblastoma multiforme 9440** (GBM).

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

- Astroblastoma **9430**
- Astrocytomas **9400** and all subtypes
  - Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401**
  - Gemistocytic astrocytoma IDH-mutant **9411**
- Diffuse midline glioma H3 K27M Mutant **9385**
- Ependymoma **9391** and all subtypes
  - Anaplastic ependymoma **9392**
  - Ependymoma, RELA fusion-positive **9396**
  - Papillary ependymoma **9393**
- Glioblastoma **9440** and all subtypes **(this is a glial tumor; however do not apply this rule to a GBM followed by a GBM)**
  - Giant cell glioblastoma **9441**
  - Glioblastoma IDH-mutant **9445**
  - Gliosarcoma **9442**
- Glioma NOS **9380**
- Oligodendroglioma and all subtypes **9450**
  - Anaplastic oligodendroglioma; IDH-mutant; 1p/19q-codeleted; IDH-mutant and 1p/19q-codeleted **9451**
- Pleomorphic xanthroastrocytoma **9424**

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

Note 3: Abstracting GBM as a new primary will allow analysis of:

- The number of tumors that recur as a more aggressive histology (GBM)
- The time interval between occurrence of the glial or astrocytic tumors and a GBM
- Which histologies are more likely to recur as a GBM

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

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

Rule M8 Abstract a **single primary**[i] when there are separate, non-contiguous tumors in the brain (multicentric/multifocal) with the **same histology XXXX**. Tumors may be any of the following combinations:

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

Example: The patient had a resection of an anaplastic astrocytoma 9401 in the right parietal lobe. Three months later the patient is diagnosed with a de novo anaplastic astrocytoma in the left parietal lobe. This is one primary because neither laterality nor timing are used to determine multiple primary status.

Note 1: **Multiple sites/subsites** and/or different **lateralities imply** either **metastatic** or **multifocal/multicentric** disease. Glioblastoma multiforme commonly exhibits multiple tumors which are described as **multifocal/multicentric**.

Note 2: Metastases are never used to determine multiple primaries. **Seeding metastasis** is often noted for the following tumors:

- Glioblastoma multiforme
- pNET-medulloblastoma
- Oligodendroglioma

Note 3: **Hereditary** syndromes frequently exhibit multiple tumors including the following:

- Neurofibromatosis type 1 (NF1)
  - Malignant peripheral nerve sheath tumors (MPNST)
- Neurofibromatosis type 2 (NF2)
  - Anaplastic ependymomas
  - Meningiomas

Note 4: Most malignant neoplasms are **single tumors** with the exception of those listed in this rule.

Note 5: This is a **change** from/**clarification** to previous rules.

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

Rule M9 Abstract **multiple primaries**[ii] when multiple tumors are present in any of the following **sites** or **subsites**:

- Any lobe of the brain **C710-C719 AND** any other part of CNS
- Cauda equina **C721 AND** any other part of CNS
- Cerebral meninges **C700 AND** spinal meninges **C701**
- Cerebral meninges **C700 AND** any other part of CNS
- Any one of the cranial nerves **C722-C725 AND** any other part of the CNS
- Any two or more of the cranial nerves
  - **C722** Olfactory, **C723** Optic, **C724** Acoustic, **C725** Cranial nerves NOS
- Meninges of cranial or peripheral nerves **C709 AND** any other part of the CNS
- Spinal cord **C720 AND** any other part of CNS
- Spinal meninges **C701 AND** any other part of CNS

Rule M10 Abstract **multiple primaries**[ii] when separate, non-contiguous tumors are two or more **different subtypes/variants** in Column 3, **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS**: Anaplastic astrocytoma IDH-mutant 9401 and gemistocytic astrocytoma IDH-mutant 9411 are both subtypes of astrocytoma NOS 9400/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Papillary ependymoma 9393 is a subtype of ependymoma NOS 9391; gliosarcoma 9442 is a subtype of glioblastoma NOS 9440. They are distinctly different histologies. Abstract multiple primaries.

Rule M11 Abstract a **single primary**[i] when separate, non-contiguous tumors are Glioma NOS and a subtype/variant of Glioma NOS.

Note: The following is a list of all tumors which would be classified as subtypes/variants of glioma NOS.

- Astroblastoma **9430**
- Astrocytomas **9400** and all subtypes
  - Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401**
  - Gemistocytic astrocytoma IDH-mutant **9411**
- Diffuse midline glioma H3 K27M Mutant **9385**
- Ependymoma **9391** and all subtypes
  - Anaplastic ependymoma **9392**
  - Ependymoma, RELA fusion-positive **9396**

  - Papillary ependymoma **9393**
- Glioblastoma **9440** and all subtypes **(this is a glial tumor; however do not apply this rule to a glial tumor followed by a GBM)**
  - Giant cell glioblastoma **9441**
  - Glioblastoma IDH-mutant **9445**
  - Gliosarcoma **9442**
- Oligodendroglioma and all subtypes **9450**
  - Anaplastic oligodendroglioma; IDH-mutant; 1p/19q-codeleted; IDH-mutant and 1p/19q-codeleted **9451**
- Pleomorphic xanthroastrocytoma **9424**:

Rule M12 Abstract a **single primary**[i] when separate, non-contiguous tumors are on **the same row** in **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3) **OR**
- A NOS histology in column 3 with an indented subtype/variant

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

Rule M13 Abstract **multiple primaries**[ii] when separate, non-contiguous tumors are on **different rows** in **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

Note 1: Each row in the table is a **distinctly different** histology.

Note 2: 8000 is considered a different row ONLY when the diagnosis is neuroepithelial tumor. If the diagnosis is cancer, NOS, do not consider 8000 to be a separate row from other histologies for the purpose of the table rules.

- Example 1: A tumor is diagnosed as 8000/3 Neuroepithelial tumor, NOS. Later, a separate tumor is diagnosed as Ependymoma 9391/3. These are considered separate rows.
- Example 2: A tumor has a provisional diagnosis of 8000/0 and further diagnosis is done. A subsequent tumor in another lobe of the brain is diagnosed as Germinoma 9064/3. These are not considered separate rows.

Rule M14 Abstract a **single primary**[i] when multiple tumors do not meet any of the above criteria.

Note: Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

This is the end of instructions for Multiple Tumors.

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[i] Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

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

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

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

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

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

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

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

This is a hierarchical list of source documentation.

1. Pathology/tissue from **resection** of primary tumor
   A. Biomarkers
      Note 1: Biomarkers are emerging as an important part of cancer diagnosis and treatment. Some biomarkers are used to determine treatment rather than histologic type. The efficacy of identification of histologic type using biomarkers differs from primary site to primary site. When a histologic type is identified using a biomarker, code the identified histology. Biomarkers do not identify all histologic types.
      Note 2: Biomarkers are not listed because they change rapidly.
      Example: Biomarkers are required to diagnose medulloblastoma SHH-activated and TP53-wildtype. Biomarkers include genetic testing, FISH, MYCN.
   B. The addendum(s) and/or comment(s)
   C. Final diagnosis / synoptic report as required by CAP
   D. CAP protocol

2. Pathology/tissue from **biopsy** of primary tumor
   A. **Biomarkers**
      Note 1: Biomarkers are emerging as an important part of cancer diagnosis and treatment. Some biomarkers are used to determine treatment rather than histologic type. The efficacy of identification of histologic type using biomarkers differs from primary site to primary site. When a histologic type is identified using a biomarker, code the identified histology. Biomarkers do not identify all histologic types.
      Note 2: Biomarkers are not listed because they change rapidly.
      Example: Biomarkers are required to diagnose medulloblastoma SHH-activated and TP53-wildtype. Biomarkers include genetic testing, FISH, MYCN.
   B. The addendum and/or comments
   C. Final diagnosis / synoptic report as required by CAP
   D. CAP protocol
   Note 1: Addendums and comments are given the second priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
   Note 2: The pathologist's diagnosis is always the most reliable, so the final diagnosis is the third priority.
   Note 3: **Do not** use the microscopic or gross section of the pathology report for coding.
   Note 4: The CAP protocol is a checklist which
      - Provides guidelines for collecting the essential data elements for complete reporting of tumors and optimal patient care.
      - Allows physicians to check multiple histologies

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

4. Tissue/pathology from a metastatic site
   Note 1: Code the behavior /3
   Note 2: The tissue from a **metastatic s**ite often shows **variations** from the primary tumor. When it is the only tissue available, it is **more accurate** than a scan.

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

6. Code the histology **documented** by the physician when none of the above are available. Use the **documentation** in the following **priority order**
   A. Treatment plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that **refers to original pathology, cytology, or scan(s)**
   D. Physician's **reference to** type of cancer (**histology**) in the medical record

   Note 1: Code the specific histology when documented.
   Note 2: Code the histology to malignant neoplasm NOS 8000 or as stated by the physician when nothing more specific is documented.

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

Coding Histology

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

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

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

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

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

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

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

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

   Example: Outpatient biopsy says probably desmoplastic medulloblastoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology desmoplastic medulloblastoma The case meets the criteria in #**3A.**

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

Example 1: The pathology diagnosis is astrocytoma consistent with gemistocytic astrocytoma IDH-mutant. The oncology consult says the patient has gemistocytic astrocytoma IDH-mutant in the occipital lobe. This is clinical confirmation of the diagnosis, code gemistocytic astrocytoma IDH-mutant. The case meets the criteria in bullet 1.

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

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

List of Ambiguous Terminology

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

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

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

Single Tumor

Rule H1 Code the **reportable CNS tumor (**Table 3 in the Equivalent Terms and Definitions) when a patient has any of the following:

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

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

Note 2: Tumors are reportable when they meet the behavior (/3) and histology requirements (see **Reportability Criteria**).

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

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

Rule H2 Code malignant meningioma **9530** when the diagnosis specifically states malignant/invasive.

Note: Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.

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

Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in **Table 3** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Table 3,** use the **ICD-O** and all **updates**.

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

Rule H4 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS such as the following:
Note: All tumors are malignant/invasive /3.

- Astrocytoma **9400** and a subtype/variant of astrocytoma
- CNS ganglioneuroblastoma **9490** and a subtype/variant of CNS ganglioneuroblastoma
- Embryonal carcinoma **9070** and a subtype/variant of embryonal carcinoma
- Ependymoma **9391** and a subtype/variant of ependymoma
- Glioblastoma **9440** and a subtype/variant of glioblastoma
- Immature teratoma **9080** and a subtype/variant of immature teratoma
- Malignant meningioma **9530** and a subtype/variant of malignant meningioma
- Malignant peripheral nerve sheath tumor **9540** and a subtype/variant of malignant peripheral nerve sheath tumor
- Medulloblastoma **9470** and a subtype/variant of medulloblastoma
- Meningeal melanoma **8720** and a subtype/variant of meningeal melanoma
- Oligodendroglioma **9450** and a subtype/variant of oligodendroglioma
- Pilocytic astrocytoma **9421** and a subtype/variant of pilocytic astrocytoma
- Pineal parenchymal tumor of intermediate differentiation **9362** and a subtype/variant of pineal parenchymal tumor of intermediate differentiation
- Sarcoma **8800** and a subtype/variant of sarcoma

Note: See **Table 3** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

This is the end of instructions for Single Tumor

Multiple Tumors Abstracted as a Single Primary

Rule H5 Code malignant meningioma **9530** when the diagnosis specifically states malignant/invasive.
Note: Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.

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

Rule H6 Code the histology when only **one** histology is present in **all** tumors.

Note 1: Use **Table 3** to code histology. New codes, terms, and synonyms are included in **Table 3** and coding errors may occur if the table is not used.

Note 2: When the histology is **not listed** in **Table 3,** use the **ICD-O** and all **updates**.

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

Rule H7 Code the **subtype/variant** when **all tumors** are a **NOS** and **a single subtype/variant** of that NOS such as the following:

Note 1: All tumors are malignant/invasive /3.

Note 2: See **Table 3** in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

- Astrocytoma **9400** and a subtype/variant of astrocytoma
- CNS ganglioneuroblastoma **9490** and a subtype/variant of CNS ganglioneuroblastoma
- Embryonal carcinoma **9070** and a subtype/variant of embryonal carcinoma
- Ependymoma **9391** and a subtype/variant of ependymoma
- Glioblastoma **9440** and a subtype/variant of glioblastoma
- Immature teratoma **9080** and a subtype/variant of immature teratoma
- Malignant meningioma **9530** and a subtype/variant of malignant meningioma
- Malignant peripheral nerve sheath tumor **9540** and a subtype/variant of malignant peripheral nerve sheath tumor
- Medulloblastoma **9470** and a subtype/variant of medulloblastoma
- Meningeal melanoma **8720** and a subtype/variant of meningeal melanoma
- Oligodendroglioma **9450** and a subtype/variant of oligodendroglioma
- Pilocytic astrocytoma **9421** and a subtype/variant of pilocytic astrocytoma
- Pineal parenchymal tumor of intermediate differentiation **9362** and a subtype/variant of pineal parenchymal tumor of intermediate differentiation
- Sarcoma **8800** and a subtype/variant of sarcoma

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

Code the histology using the rule that fits the case.

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

Introduction

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

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

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

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

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

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

- For cases diagnosed prior to 1/1/2023, these neoplasms are reportable in North American as malignant 9421/3 for all CNS sites with the exception of the optic nerve:
  - WHO Classification Tumors of the Central Nervous System and IARC designate pilocytic astrocytoma as a synonym for optic glioma
  - When the primary site is optic nerve and the diagnosis is either optic glioma or pilocytic astrocytoma, the behavior is non-malignant and coded 9421/1
  - Beginning with cases diagnosed 1/1/2023 forward, pilocytic astrocytoma/juvenile pilocytic astrocytoma are to be reported as 9421/1 for ***all*** CNS sites.

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

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

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

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Equivalent or Equal Terms

These terms can be used interchangeably:

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

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Terms that are NOT Equivalent or Equal

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

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

Changes from 2007 MPH Rules

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

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

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

New for 2023

Beginning with cases diagnosed 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma will **no longer be reported as malignant (9421/3)**. These neoplasms will continue to be reportable as a benign CNS tumor with a behavior of /1. Historically, pilocytic astrocytomas were coded as 9421/3 from 1976-2000. Beginning in 2001 with the release of ICD-O-3, WHO changed the behavior to /1, however, the standard setters opted to continue collecting these cases using malignant /3 behavior. This practice will continue through 12/31/2022.

Reportability Criteria for Non-Malignant CNS Neoplasms

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

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

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Directory of Sections and Tables

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

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Section 1: Behavior Code

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

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

Priority Order for Using Documentation to Assign Behavior

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

D. Angiogram

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

Table 1: WHO Grades of Select CNS Neoplasms

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

WHO Grade Definitions

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

WHO Grade II CNS Tumors: Non-malignant and Malignant

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

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

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

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

Table Instructions

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

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

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

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

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

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Section 2: Reportable Primary Sites and Histologies

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

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

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

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

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Priorities for Coding Primary Site

Note 1: **Always** check the **operative report(s)** which will have information on whether the surgery or biopsy was **intracranial** (inside the cranium/skull) or **intraspinal** (within the dura/meninges covering the spinal cord)

Note 2: Code **the specific primary site**. Use an **NOS** site code **only** when a specific site is not known.

Note 3: See **Table 2**: **Reportable Primary Sites** to confirm the primary site is reportable.

Note 4: When the primary site is cranial nerve **OR** cranial nerve meninges, see **Table 3**: **Reportability of Non-Malignant Cranial Nerve (CN) Tumors** to confirm the site of the tumor is intracranial (reportable) or extracranial (not reportable)

Note 5: See **Table 4**: **Non-Reportable Neoplasms** for site/histology combinations and histologies that are not reportable.

Note 6: When the primary site is brain or intracranial glands, see **Table 5**: **Histologic Types of Non-Malignant Intracranial (Brain and Gland) Tumors** to confirm site/histology combinations.

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

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

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Reportable Primary Site Groups

The two major groups of reportable sites are:

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

Reportable Primary Sites and their ICD-O Codes

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

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Table 2: Reportable Primary Sites

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

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

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Site Group	Reportable Subsite Terms and Code
Ill-Defined Sites Central Nervous System	Nervous system NOS **C729** Overlapping lesion of brain and central nervous system **C728**
Intracranial Duct and Glands	Craniopharyngeal duct **C752** Pineal gland **C753** Pituitary gland **C751**
Meninges	Cerebral meninges **C700** Meninges NOS **C709** Spinal meninges **C701**
Spinal Sites	Cauda equina **C721** Conus medullaris/filum terminale **C720** Meninges NOS **C709** Spinal cord **C720** Spinal meninges **C701**

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Table 3: Reportability of Non-Malignant Cranial Nerve (CN) Tumors

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

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

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

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

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Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Trochlear CN 4 **C725**	Superior orbital fissure	Arises from the **dorsal brain stem**, loops around the brainstem and passes anteriorly within the **subarachnoid space**. It travels between the **superior cerebellar and posterior cerebral arteries** and through the **dura,** enters **cavernous sinus.**	Enters the **orbital fissure**.
Trigeminal CN 5 **C725** ***Note:*** Trigeminal is derived from Latin trigeminus which means **born in threes** (tri) and born **at the same time** (germinal). As the name implies, the nerve **separates** into **three branches**; ophthalmic, maxillary, and mandibular	The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina, the superior orbital fissure, the foramen rotundum and the foramen ovale.	CN5 originates in the **pons**. Upon leaving the pons it enters a **small fossa** posterior and inferolateral to the cavernous sinus called **Meckel's (trigeminal) cave**.	• **Ophthalmic nerve branch** crosses the **pterygopalatine fossa**, inclines laterally on the back of the **maxilla**, and enters the **orbit** through the inferior orbital fissure where it is called the **infraorbital** nerve. It ends beneath the **quadatus labii superius**, and divides into multiple branches that spread to the side of the **nose**, the lower **eyelid**, and the upper **lip** • **Maxillary** nerve leaves foramen rotundum and traverses the **infraorbital groove** and **canal** in the floor of the **orbit**, and appears on the face at the infraorbital foramen. • **Mandibular** nerve leaves via the foremen ovale travels along the **mandibular** groove

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Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Abducent CN 6 **C725**	Cranial meninges	Exits brainstem at junction of **pons** and the **medulla**, enters the **subarachnoid** space and runs upward between the pons and the **clivus** entering the **cavernous sinus.**	**Dorello's canal** and travels to the tip of the **temporal bone**. Enters **orbit**
Facial CN 7 **C725**	Internal acoustic meatus	**CN7** originates in the **pons,** along the posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity)	Enters the **temple** through the **internal auditory meatus** and runs through the **facial canal**.
Acoustic or vestibulocochlear CN 8 **C724**	Internal acoustic meatus	Originates in the **brain stem** (**medulla oblongata**) **between** the base of the brain (**pons**) and the **spinal cord** Both the **vestibular** branch and the **cochlear** branch are located in the **inner ear**	
Glossopharyngeal CN 9 **C725**	Jugular foramen	**Originates** in the anterior portion of the **medulla oblongata**	**Jugular foramen** Between the i**nternal jugular** vein and **internal carotid artery** Lies on the **stylopharyngeus** and **middle pharyngeal constrictor muscle** Passes under the **hypoglossus muscle** **Palatine tonsil** Extends to **mucous glands** of the **mouth**, and **base** of the **tongue**

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

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

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

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

Table 4: Non-Reportable Neoplasms

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

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

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

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

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

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

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

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

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

Use **Table 5** to:

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

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

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

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

Histology Term and Code	Most Common Intracranial Primary Site
Dysplastic gangliocytoma 9493/0	Cerebellum **C716**
Juvenile xanthogranuloma 9749/1	Intraventricular **C715**
Meningioma (rare) 9530/0	Intraventricular **C715**
Myxopapillary ependymoma 9394/1	4th ventricle **C717**
Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1	**Optic nerve C723**
Pineocytoma 9361/1	Pineal gland C**753**
Pituicytoma 9432/1*	Pituitary gland **C751**, sella turcica **C751,** suprasellar **C719**
Pituitary adenoma 8272/0	Pituitary gland **C751**
Prolactinoma 8271/0	Pituitary gland **C751**
Subependymal giant cell tumor (SEGA) 9384/1	Lateral ventricles **C715**
Subependymoma 9383/1	Intraventricular site (lateral/third ventricle, rare **C715** and IV ventricle **C717**)

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

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

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

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

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

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

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

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

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Diffuse astrocytoma, MYB- or MYBL1 altered 9421/1 *Note 1:* Beginning 1/1/2023, diffuse astrocytoma, MYB- or MYBL1 altered is the preferred term for 9421/1. *Note 2:* Beginning 1/1/2023, pilocytic astrocytoma/juvenile pilocytic astrocytoma are coded 9421/1. Cases diagnosed prior to 1/1/2023 are coded 9421/3.	Angiocentric glioma Diffuse low-grade glioma, MAPK pathway-altered Juvenile pilocytic astrocytoma Pilocytic astrocytoma	
Dysembryoplastic neuroepithelial tumor 9413/0 *Note:* DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in column 1.	DNET	
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	Cavernoma	Cavernous hemangioma **9121/0** Venous hemangioma **9122/0**
Juvenile xanthogranuloma 9749/1		
Leiomyoma 8890/0		
Lipoma 8850/0		Hibernoma **8880/0**
Meningeal melanocytosis 8728/0		Meningeal melanocytoma **8728/1**

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

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Meningioma 9530/0	Lymphoplasmacyte-rich meningioma Metaplastic meningioma Microcystic meningioma Secretory meningioma	Angiomatous meningioma **9534/0** Atypical meningioma **9539/1** Clear cell/chordoid meningioma **9538/1** Fibrous meningioma **9532/0** Meningothelial meningioma **9531/0** Psammomatous meningioma **9533/0** Transitional meningioma **9537/0**
Multinodular and vacuolating neuronal tumor 9509/0 ***Note:*** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on separate rows in column 1.	MVNT	
Myofibroblastoma 8825/0		Inflammatory myofibroblastic tumor **8825/1**
Myxopapillary ependymoma 9394/1		
Neurocytoma 9506/1	Central neurocytoma Cerebellar liponeurocytoma Extraventriculare neurocytoma Lipomatous medulloblastoma Medullocytoma Neurolipocytoma	

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

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Neuroepithelial tumor, benign 8000/0 *Note*: Neuroepithelial tumor is a rare tumor specific to children. These neoplasms have numerous subtypes which are not easily identified so a specific type may not be provided in the pathology report. WHO has not proposed an ICD-O code for this entity. The current option is to assign code 8000. Because these tumors are different, they are on a separate row.	Neuroepithelial tumor, NOS 8000/1	
Neurofibroma 9540/0	Atypical neurofibroma	Plexiform neurofibroma **9550/0**
Optic glioma/pilocytic astrocytoma 9421/1		
Papillary glioneuronal tumor 9509/1 ***Note 1:*** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on rows in column 1. ***Note 2:*** Beginning with cases diagnosed 1/1/2023 forward, leptomeningeal glioneuronal tumor is coded 9509/3. See the Malignant CNS rules.	Diffuse leptomeningeal glioneuronal tumor (see note 2) Rosette-forming glioneuronal tumor	
Paraganglioma 8693/1		
Perineurioma 9571/0		
Pineocytoma 9361/1		
Pituicytoma 9432/1*		

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

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Pituitary adenoma 8272/0	Corticotroph Gonadotroph adenoma Somatotroph adenoma Thyrotroph adenoma Null cell adenoma Plurihormonal and double adenomas	
Polymorphous low-grade neuroepithelial tumor of the young 9413/0 ***Note*:** DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in column 1.	PLNTY	
Prolactinoma 8271/0		
Rhabdomyoma 8900/0		
Schwannoma 9560/0	Acoustic neuroma Cellular schwannoma Neurilemoma Neurinoma Plexiform schwannoma	Melanotic schwannoma **9560/1***
Solitary fibrous tumor Grade 1 8815/0	Hemangiopericytoma Grade 1	Solitary fibrous tumor/hemangiopericytoma Grade 2 **8815/1***
Spindle cell oncocytoma 8290/0		
Subependymal giant cell astrocytoma 9384/1		
Subependymoma 9383/1		
Teratoma 9080/1		

Section 3: Additional Information to Complete Abstract

Conflicting Information on Pathology Report(s)

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

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

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

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

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

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

Table 7: Paired Sites

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

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

Note 1: Midline tumors are most common for the following histologies: glioblastoma multiforme, meningiomas, lymphomas (usually located near the midline), and epidermoid cysts (which can cross the midline via the subarachnoid space).
Note 2: SEER allows laterality to be coded for sites other than those in the table.

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

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

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

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

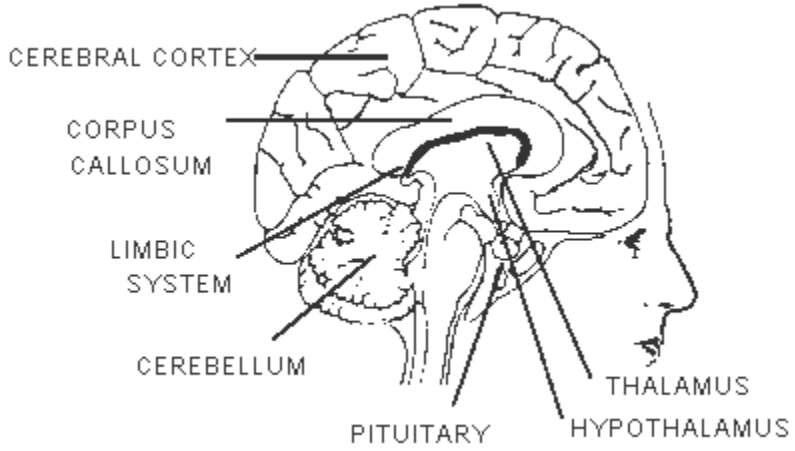
The table identifies non-malignant tumors that have the **potential** of **transforming to** a malignant tumor (**new primary**).

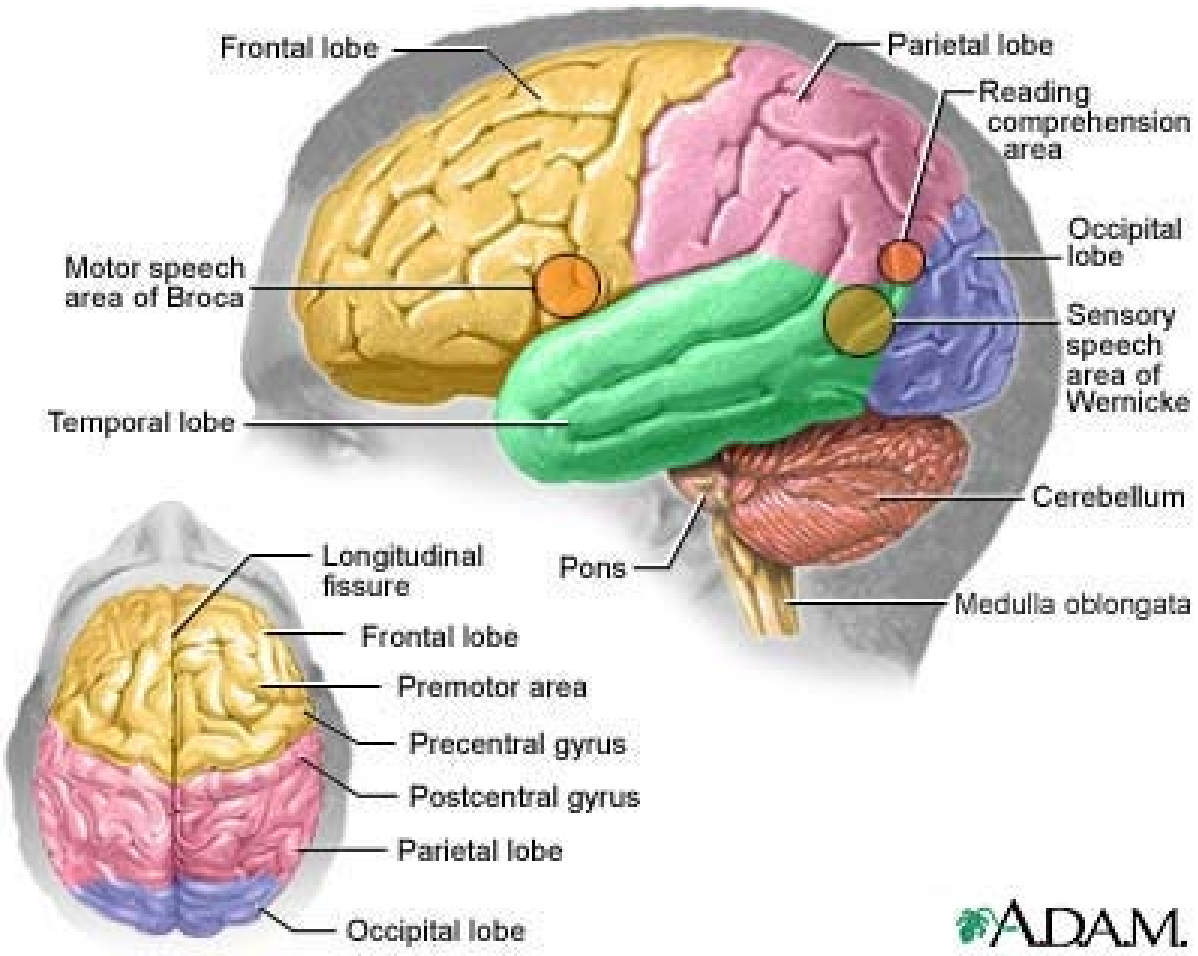
Column 1 is the **non-malignant** ICD-O histology term and code.
Column 2 is the **malignant** /3 ICD-O histology term and code to which the non-malignant tumor can transform.

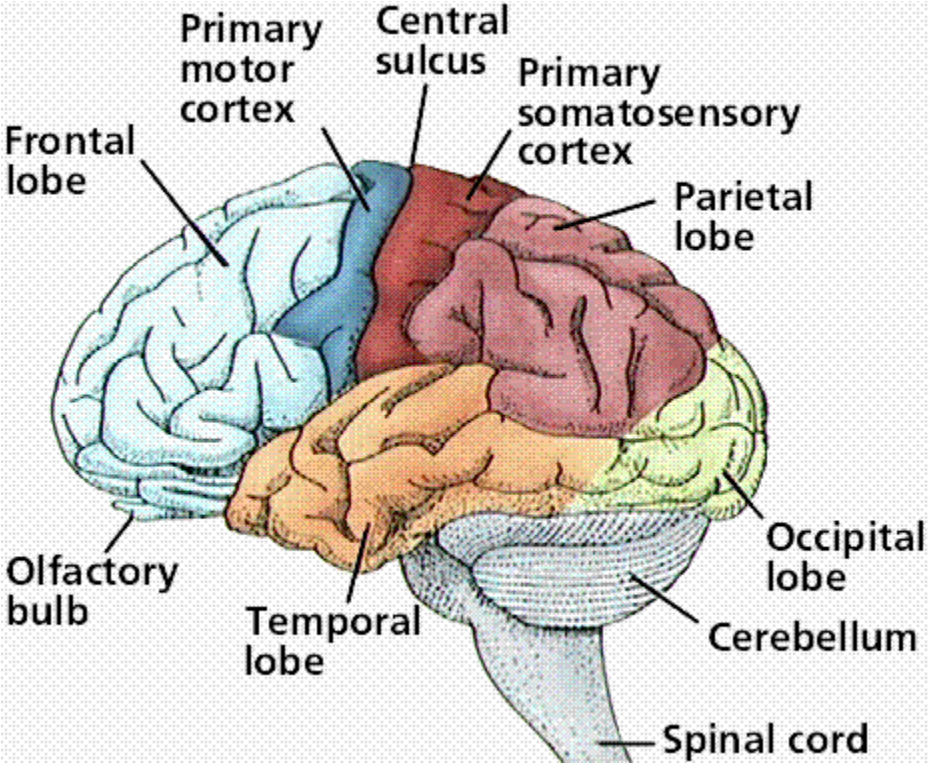
Original Histology and Code	Transformed Histology and Code
Chondroma **9220/0**	Chondrosarcoma **9220/3**
Ganglioglioma **9505/1**	Anaplastic ganglioglioma **9505/3**
Hemangioma **9120/0**	Angiosarcoma **9120/3**
Hemangiopericytoma **9150/1**	Anaplastic hemangiopericytoma **9150/3**
Leiomyoma **8890/0**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

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

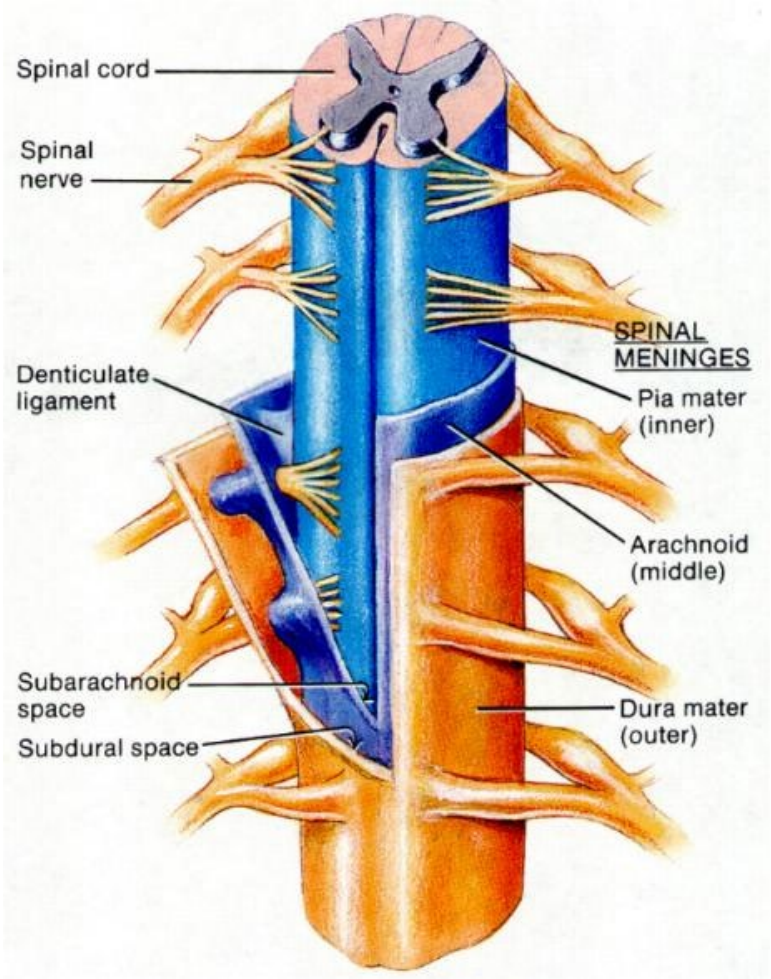
Illustrations



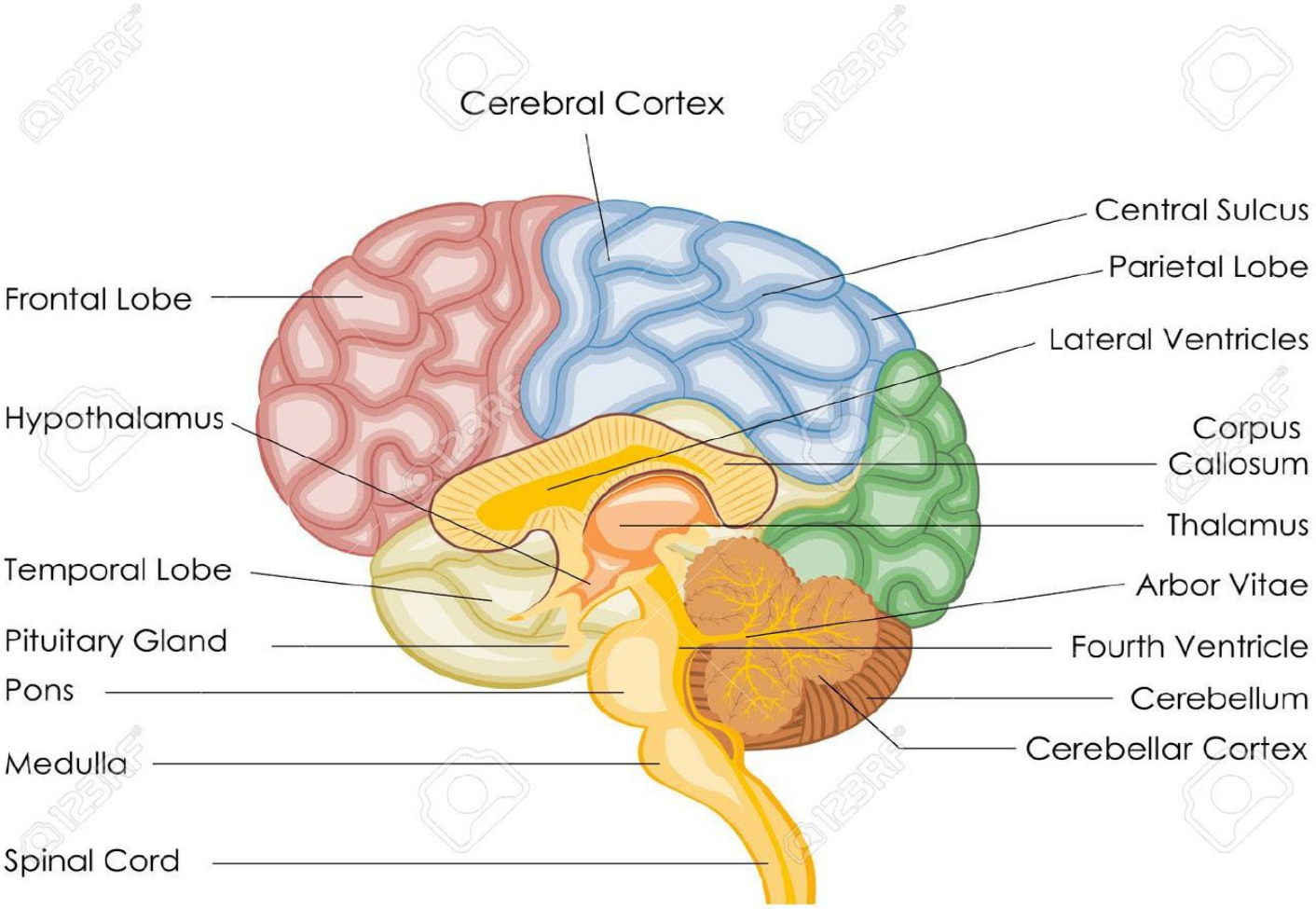




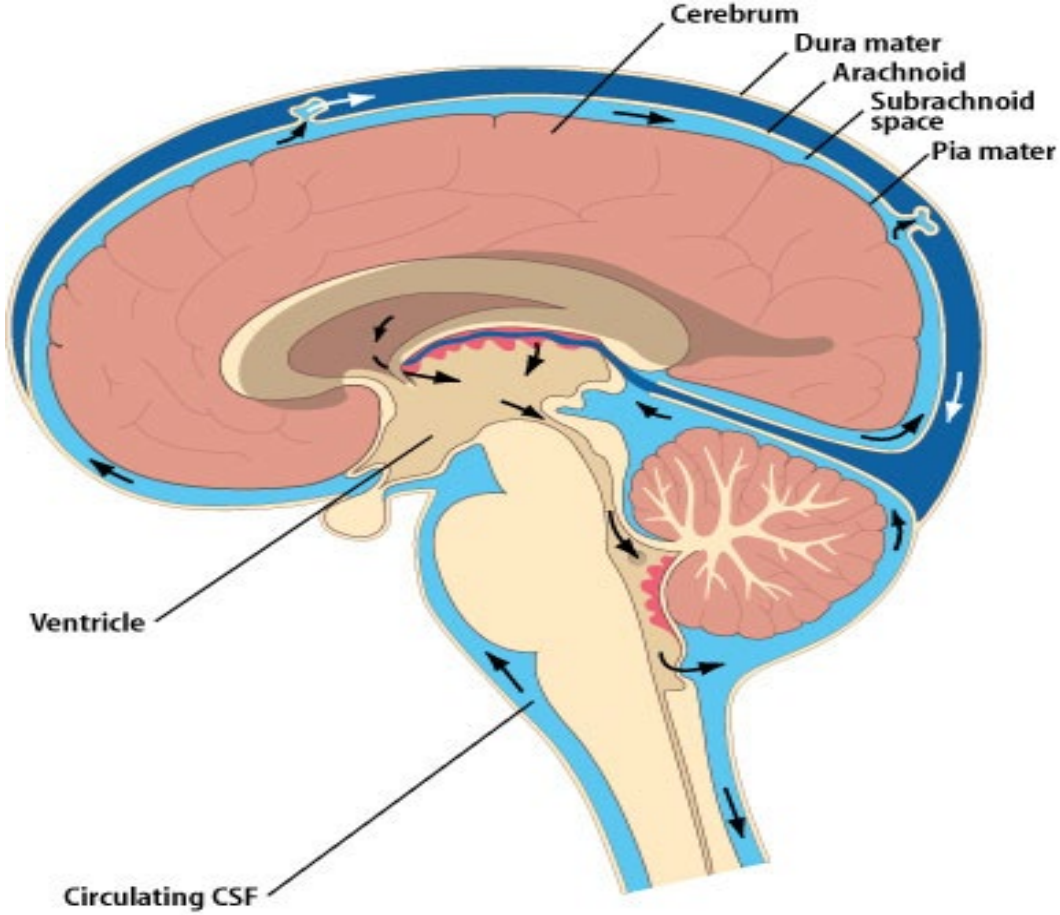
Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)



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



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



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

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

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

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted
Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
- Outpatient biopsy with no follow-up information available
- Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors.

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

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

Single Tumor

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

Rule M2 Abstract a **single primary**[i] when there is a **single tumor.**

Note 1: A single tumor is **always** a single primary.
Note 2: The tumor may overlap onto or extend into an adjacent/contiguous site or subsite.
Note 3: The tumor may have multiple histologic components.
Note 4: The tumor may be diagnosed as borderline /1 on scans and subsequent pathology proves a benign /0 tumor.
Example: Patient has a MRI with a diagnosis of a benign /0 tumor. The pathology report from the subsequent surgery shows a borderline /1 tumor. This is a single tumor and **a single primary**. (It is the same tumor as seen on the MRI; better information is provided by the pathology report.)

Rule M3 Abstract a **single primary**[i] (the malignant) when a single tumor meets the following two criteria:

1. The original diagnosis was non-malignant /0 or /1 **AND**
   - First course treatment was active surveillance (no tumor resection). Diagnosis was:
     - Clinical
     - Radiographic
     - Stereotactic biopsy

2. Subsequent resection pathology is malignant /3

Note 1: This is a **new rule** which **clarifies** that a single tumor is **always** a single primary and the malignant behavior is reported.
Note 2: Use the Malignant CNS and Peripheral Nerves Rules to code histology.
Note 3: The **resection pathology** is **more accurate** than clinical, radiographic or stereotactic biopsy information. While stereotactic biopsy provides a pathologic specimen, it is small and may not have included the malignant portion of tumor.
Note 4: There is **no time requirement** from initial diagnosis to resection.
Note 5: Edit the original abstract as follows:
- Do **not** change **date of diagnosis**.
- For cases which have been abstracted, **change behavior** code on original abstract from /0 or /1 to /3.
- **Report** all data changes for cases which have been submitted to the central registry.

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

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

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

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

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

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

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

Note 2: This is a single tumor; single primary

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

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

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

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

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

This is the end of instructions for Single Tumor.

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

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

Multiple Tumors

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Rule M10 Abstract a **single primary**[i] when there are separate/non-contiguous tumors in the brain (multicentric/multifocal) with the **same histology XXXX**. Tumors may be in any of the following locations and/or lateralities:

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

***Exception 1:** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on separate rows in Table 6.

***Exception 2:** DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in Table 6.

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

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

Note: The same row means the tumors are:

- The same histology* (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3). NOS and subtype/variants are:
  - Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
  - Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
  - Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
  - Lipoma **8850/0** and a subtype/variant of lipoma
  - Meningeal melanocytosis **8728/0** and a subtype/variant of meningeal melanocytosis
  - Meningioma **9530/0** and a subtype/variant of meningioma

- Myofibroblastoma **8825/0** and a subtype/variant of myofibroblastoma
- Neurofibroma **9540/0** and a subtype/variant of neurofibroma
- Schwannoma **9560/0** and a subtype/variant of schwannoma
- Solitary fibrous tumor WHO Grade 1 **8815/0** and a subtype/variant of solitary fibrous tumor WHO Grade 1

***Exception 1:** MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on separate rows in Table 6.

***Exception 2:** DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in Table 6.

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

Note 1: Each row in the table is a **distinctly different** histology.

Note 2: MVNT and papillary glioneuronal tumor have the same ICD-O code but are distinctly different histologies based on behavior codes. Because they are different, they are on different rows in Table 6.

Note 3: DNET and PLNTY have the same ICD-O code but are distinctly different histologies. Because they are different, they are on separate rows in Table 6.

Note 4: 8000 is considered a different row ONLY when the diagnosis is neuroepithelial tumor. If the diagnosis is cancer, NOS, do not consider 8000 to be a separate row from other histologies for the purpose of the table rules.

- Example 1: A tumor is diagnosed as 8000/1 Neuroepithelial tumor, NOS. Later, a separate tumor is diagnosed as Hemangioma 9120/0. These are considered separate rows.
- Example 2: A tumor has a provisional diagnosis of 8000/0 and further diagnosis is done. A subsequent tumor in another lobe of the brain is diagnosed as myofibroblastoma 8825/0. These are not considered separate rows.

Rule M13 Abstract a **single primary**[i] when the tumors do not meet any of the above criteria.

Note: These rules are hierarchical. Use this rule **ONLY** when the previous rules do not apply.

This is the end of instructions for Multiple Tumors

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[i] Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

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

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

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

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

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

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

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

This is a hierarchical list of source documentation:

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

- Biomarkers are not listed because they change rapidly.

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

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

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

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

Note 4: The CAP protocol is a checklist which

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

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

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

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

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

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

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

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

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

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

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

Coding Histology

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

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

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

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

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

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

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

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

Example: Outpatient biopsy says probably central neurocytoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology central neurocytoma. The case meets the criteria in #**3A.**

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

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

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

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

List of Ambiguous Terminology

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

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

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

Single Tumor

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

This is the end of instructions for Single Tumor.

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

Multiple Tumors Abstracted as a Single Primary

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

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

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

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

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

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

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

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

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

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

Note 1: Only report tumors such as:

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Introduction

Note 1: The group name "urinary sites" include: Renal pelvis C659; ureter C669; trigone of bladder C670; dome of bladder C671; lateral wall of bladder C672; anterior wall of bladder C673; posterior wall of bladder C674; bladder neck C675; ureteric orifice C676; urachus C677; overlapping lesion of bladder C678; bladder NOS C679; urethra C680; paraurethral gland C681; overlapping lesion of urinary organs C688; and urinary system NOS C689.

Note 2: Tables and rules refer to **ICD-O** rather than **ICD-O-3**. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

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

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

Note 4: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

In US, 90% of bladder tumors are urothelial carcinoma; less than 5% are pure squamous cell carcinoma or pure adenocarcinoma.

Urothelial carcinoma originates in urothelial/transitional cells which line the urethra, bladder, ureters, and renal pelvis and has two major subdivisions: papillary and non-papillary.

- Papillary carcinoma: (commonly in bladder, ureter, or renal pelvis): A warty growth which projects from the wall on a stalk
  - Non-invasive papillary urothelial carcinoma (occasionally called in situ)
  - Invasive papillary urothelial carcinoma
- Non-papillary urothelial: originates within the mucosa and does not project from the wall
  - Non-invasive carcinoma in situ (CIS)
  - Invasive urothelial carcinoma

Note: Both urothelial carcinoma and papillary urothelial carcinoma can be in situ /2 or invasive /3. Code the behavior specified in the pathology report.

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multifocal/Multicentric Tumors of Urinary Sites

Multifocality of urothelial carcinoma is a common finding. The phenomenon of multiple tumors has been theorized as being a result of the field effect.

The field effect concept has two main theories:

1. **Monoclonal**: A single malignant cell spreads throughout the urothelium by:
   a. Intraluminal spread with secondary implantation in different sites within the urinary tract **OR**
   b. Intraepithelial migration
2. **Oligoclonal**: Multifocal/multicentric tumors develop secondary to a field effect precipitated by carcinogens. The carcinogens cause genetic alterations at different sites within the urinary tract.

Neither theory has been conclusively proven.

Flat/urothelial carcinoma in situ can have a widespread effect as a result of direct spread of neoplastic cells within the epithelium.

The rules for coding histology and defining the number of primaries are an attempt to reconcile these observations in order to provide **incidence** data that are consistent and reproducible.

Changes from 2007 MPH Rules

1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, 2018 Rules instruct "Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor)."
2. There are no significant changes in histology terms or codes in the 2016 WHO edition.

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**. Urothelial carcinoma **and** small cell neuroendocrine carcinoma is **equivalent** to urothelial carcinoma **with** small cell neuroendocrine carcinoma.
- Flat transitional cell carcinoma; flat urothelial carcinoma; urothelial carcinoma in situ; noninvasive flat carcinoma; in situ transitional cell carcinoma
- Multifocal; multicentric
- Noninvasive may describe either in situ papillary carcinoma or flat urothelial cell carcinoma
- Papillary transitional cell carcinoma; papillary urothelial carcinoma
- Simultaneous; synchronous; existing at the same time; concurrent; prior to first course treatment
- Topography; site code
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a standard manner in clinical diagnoses, scans, or consults. **Disregard** the terms **unless** there is a physician's statement that the term is malignant/cancer
  - These terms are used **ONLY** to determine multiple primaries
  - **Do not** use these terms for casefinding or for determining reportability
- Type; subtype; variant
- Urothelial carcinoma; transitional cell carcinoma
- Urothelium; epithelium; transitional epithelium

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
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Terms that are Not Equivalent or Equal

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

- Carcinoma, NOS (8010) and adenocarcinoma, NOS (8140) are not equivalent
- **Phenotype** is not equivalent to **subtype/type/variant**
- **Noninvasive, papillary urothelial carcinoma, flat urothelial carcinoma** are not equivalent

  Note: Noninvasive is **not equivalent** to either **papillary** urothelial or **flat** urothelial carcinoma. **Both Ta** and **Tis** tumors are technically noninvasive. Code the histology specified by the **pathologist.**
- **Papillary growth pattern** is not equivalent to **papillary urothelial carcinoma**

Instructions for Coding Primary Site

The following instructions are in priority order.

1. Code overlapping lesion of urinary bladder **C678** when:
   A. A single tumor of any histology overlaps subsites of the bladder
   B. A single tumor or non-contiguous tumors which are:
      - **Urothelial carcinoma in situ 8120/2 AND**
      - Involves only bladder and one or both ureters (no other urinary sites involved)

   ***Note*:** Overlapping non-invasive tumors of the bladder and ureter almost always originate in the bladder. They extend/overlap into the ureter by spreading along the mucosa. It is important to code these primaries to bladder C678, NOT to overlapping lesion of urinary organs C688.
2. Code bladder NOS **C679** when there are **multiple non-contiguous tumors** within the **bladder AND** the subsite/origin is unknown/not documented.
3. Code overlapping lesion of urinary organs **C688** when a single tumor overlaps two urinary sites and the origin is unknown/not documented.
   Note: See the following examples of contiguous urinary sites where overlapping tumor could occur:
   - Renal pelvis and ureter
   - Bladder and urethra

- Bladder and ureter (for all histologies other than in situ urothelial cell)

4. Code Urinary System NOS **C689** when there are **multiple non-contiguous tumors** in **multiple organs** within the urinary system.
   Note: The physician subject matter experts (SME) discussed the issue of coding primary site for **multifocal/multicentric** urinary tract carcinoma. Although the SMEs understood and acknowledged the importance of coding a specific primary site, there is **no literature** or **criteria** for **determining** the organ of **origin** for multiple tumors involving multiple urinary sites.

Table 1: ICD-O Primary Site Codes

Use the following table to determine the correct site code.

Column 1 contains the site term and ICD-O code.
Column 2 contains synonyms for the site code and term in column 1.

Site Term and code	Synonyms
Bladder, anterior wall **C673**	-
Bladder, dome **C671**	Roof Vault Vertex
Bladder, lateral wall **C672**	Lateral to ureteral orifice Left wall Right wall Sidewall
Bladder neck **C675**	Internal urethral orifice Vesical neck
Bladder NOS **C679**	Lateral posterior wall (**no hyphen**)
Bladder, overlapping lesion **C678**	Fundus Lateral-posterior wall (**hyphen**)
Bladder, posterior wall **C674**	-

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Site Term and code	Synonyms
Bladder, trigone **C670**	Base of bladder Below interureteric crest Below interureteric field Below interureteric ridge Floor of bladder
Bladder, urachus **C677**	Mid umbilical ligament Urachal remnant
Bladder, ureteric orifice **C676**	Just above ureteric orifice
Overlapping lesion of urinary organs **C688**	-
Paraurethral gland **C681**	-
Renal pelvis **C659**	Pelvis of kidney Pelviureteric junction Renal calyces Renal calyx
Ureter **C669**	-
Urethra **C680**	Cowper gland Littre glands Prostatic utricle Urethral gland
Urinary system NOS **C689**	-

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Table 2: Specific Histologies, NOS, and Subtypes/Variants

Use Table 2 as directed by the **Histology Rules** to assign the more common histology codes for urinary tract neoplasms.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including rhabdomyosarcoma 8900/3 (column 3). Rhabdomyosarcoma is also a NOS because it has a subtype/variant 8910/3. The subtype/variant is indented under the NOS (rhabdomyosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that rhabdomyosarcoma has a subtype/variant.

When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, rhabdomyosarcoma **8900/3** and embryonal rhabdomyosarcoma **8910/3** are a NOS and a subtype/variant, **NOT** two different subtypes.

Table begins on next page

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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Adenocarcinoma NOS **8140** ***Note***: Urachal **carcinoma** NOS is coded 8010/3. Urachal **adenocarcinoma** is coded 8140/3.	Mixed adenocarcinoma Urachal adenocarcinoma	Clear cell carcinoma **8310** Endometrioid carcinoma **8380** Enteric adenocarcinoma **8144** Mucinous adenocarcinoma **8480**
Malignant melanoma **8720/3**		
Malignant perivascular epithelioid cell tumor **8714/3**	Malignant PEComa	
Neuroendocrine carcinoma, NOS **8246/3**		Large cell neuroendocrine tumor/combined large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor, NOS **8240/3**	Neuroendocrine tumor, grade 1 Well differentiated neuroendocrine carcinoma Neuroendocrine carcinoma, low grade	Neuroendocrine tumor, grade 2 **8249/3**
Sarcoma NOS **8800/3** ***Note:*** Rhabdomyosarcoma **8900** is a NOS with a subtype/variant of embryonal rhabdomyosarcoma/sarcoma botryoides **8910/3.**		Angiosarcoma **9120/3** Chondrosarcoma **9220/3** Leiomyosarcoma **8890/3** Liposarcoma **8850/3** Malignant peripheral nerve sheath tumor (MPNST) **9540/3** Pleomorphic sarcoma **8802/3** Rhabdomyosarcoma **8900/3** Embryonal rhabdomyosarcoma/sarcoma botryoides **8910/3**

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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Squamous cell carcinoma **8070**	Pure squamous cell carcinoma SCC Pure squamous carcinoma of urothelial tract	Verrucous carcinoma **8051**
Urothelial carcinoma **8120** ***Note 1:*** Previously called **transitional cell** carcinoma, a term that is no longer recommended. ***Note 2:*** Micropapillary **8131** is a subtype/variant of papillary urothelial carcinoma **8130**. It is an invasive /3 neoplasm with aggressive behavior. ***Note 3:*** The histology term is exactly Plasmacytoid/signet ring cell/diffuse variant. All three terms are used together to indicate a specific variant (coded 8082/3).	Clear cell (glycogen-rich) urothelial carcinoma **8120/3** Conventional urothelial carcinoma **8120/3** Infiltrating urothelial carcinoma **8120/3** Infiltrating urothelial carcinoma with divergent differentiation **8120/3** Infiltrating urothelial carcinoma with endodermal sinus lines **8120/3** Infiltrating urothelial carcinoma with glandular differentiation **8120/3** Infiltrating urothelial carcinoma with squamous differentiation **8120/3** Infiltrating urothelial carcinoma with trophoblastic differentiation **8120/3** Large nested urothelial carcinoma **8120/3**	Giant cell urothelial carcinoma **8031/3** Lymphoepithelioma-like urothelial carcinoma **8082/3** Plasmacytoid/signet ring cell/diffuse variant (see Note 3) Papillary urothelial (transitional cell) carcinoma in situ **8130/2** invasive **8130/3** low-grade papillary urothelial carcinoma with inverted growth pattern **8130/2** non-invasive papillary urothelial carcinoma, high-grade **8130/2** non-invasive papillary urothelial carcinoma, low-grade **8130/2** Micropapillary urothelial carcinoma **8131/3** Plasmacytoid urothelial carcinoma/ sarcomatoid urothelial carcinoma **8122/3** Poorly differentiated carcinoma/poorly differentiated urachal carcinoma **8020/3**

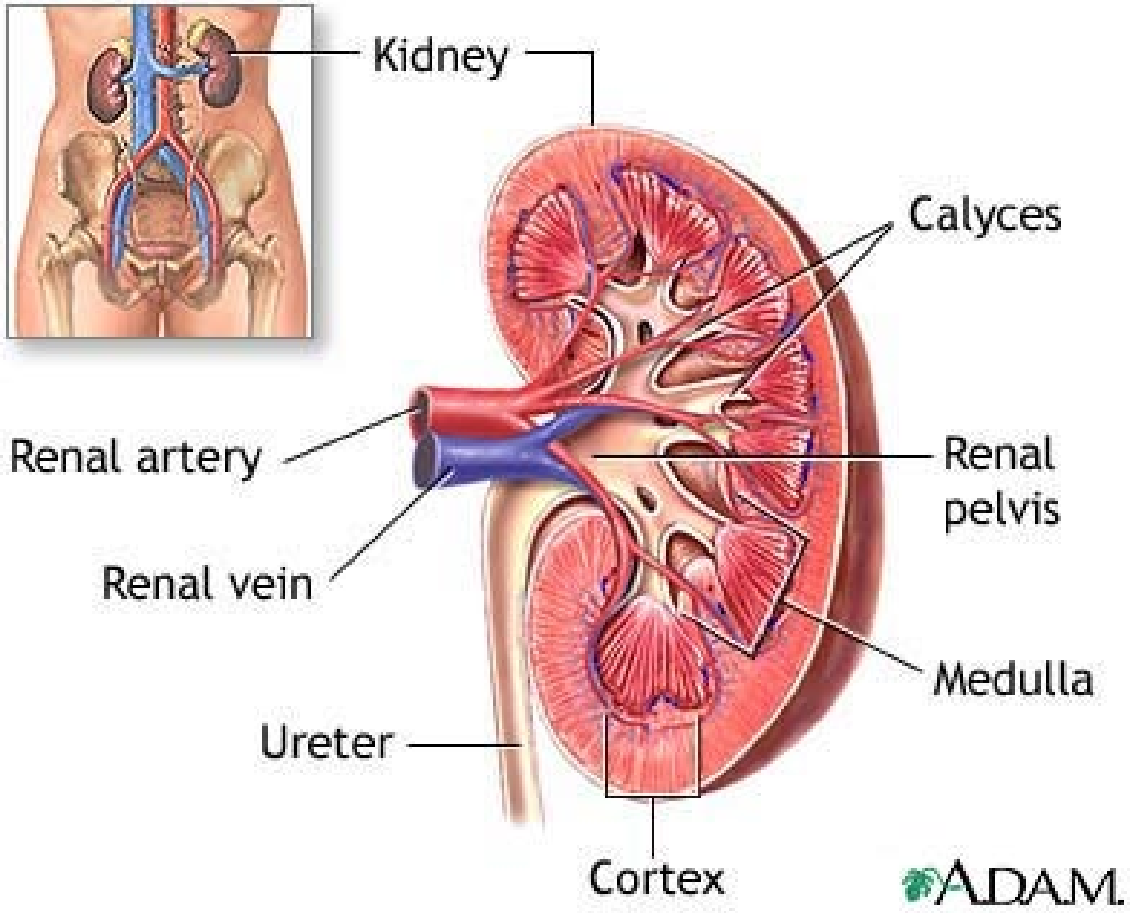
Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
	Lipid-rich urothelial carcinoma **8120/3** Microcystic urothelial carcinoma **8120/3** Nested urothelial carcinoma **8120/3** Plasmacytoid urothelial carcinoma **8120/3** Tubular and microcystic urothelial carcinoma **8120/3** Urothelial carcinoma in situ **8120/2**	

Table 3: Non-Reportable Urinary Tumors

Column 1 contains the terms and codes (if applicable) for the non-reportable histology.
Column 2 contains **synonyms** of the histology term in column 1. Synonyms have the **same code** as the term in Column 1.

Histology Term and Code	Synonyms
Benign perivascular epithelioid cell tumor **8714/0**	Benign PEComa
Granular cell tumor **9580/0**	
Hemangioma **9120/0**	
Inflammatory myofibroblastic tumor **8825/1**	
Inverted urothelial papilloma **8121/0**	
Leiomyoma **8890/0**	
Melanosis **No code**	
Neurofibroma **9540/0**	
Nevus **8720/0**	
Papillary urothelial neoplasm of low-malignant potential **8130/1**	
Paraganglioma **8693/1**	Extra-adrenal pheochromocytoma
Solitary fibrous tumor **8815/1**	
Squamous cell papilloma **8052/0**	Keratotic papilloma
Urothelial dysplasia **No code**	
Urothelial papilloma **8120/0**	
Villous adenoma **8261/0**	

Illustrations



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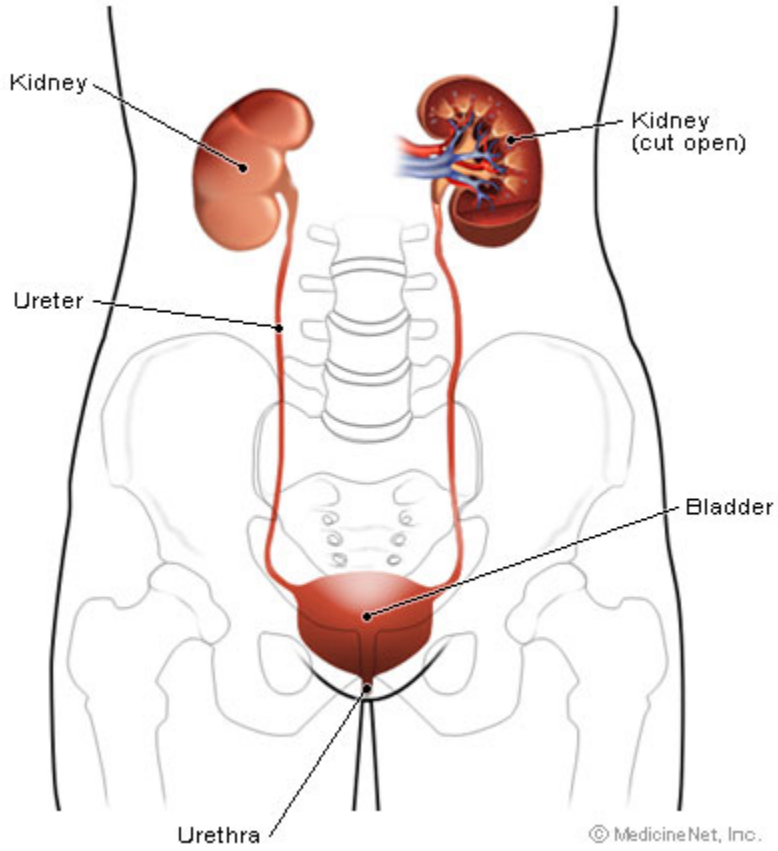

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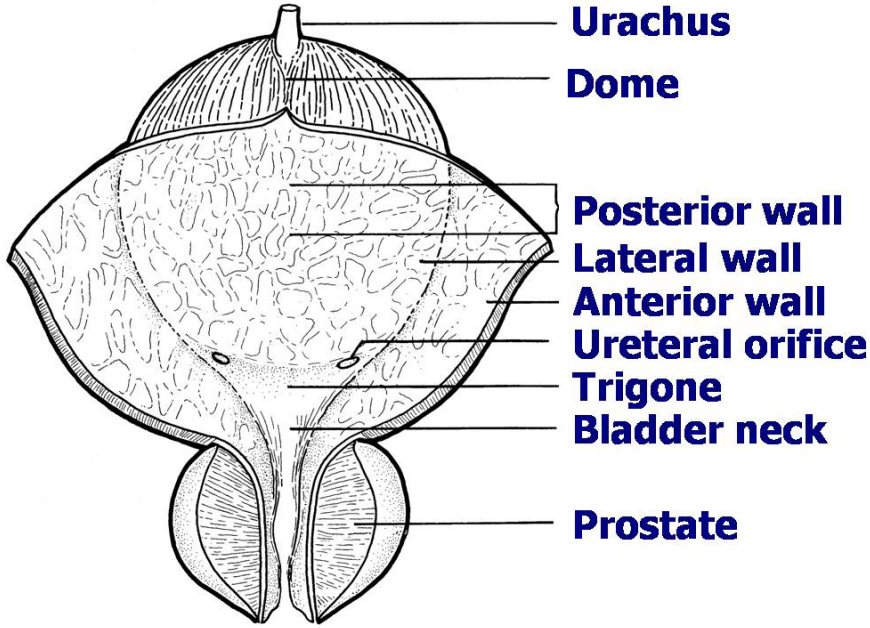

Source: TNM Atlas, 3rd edition, 2nd revision

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
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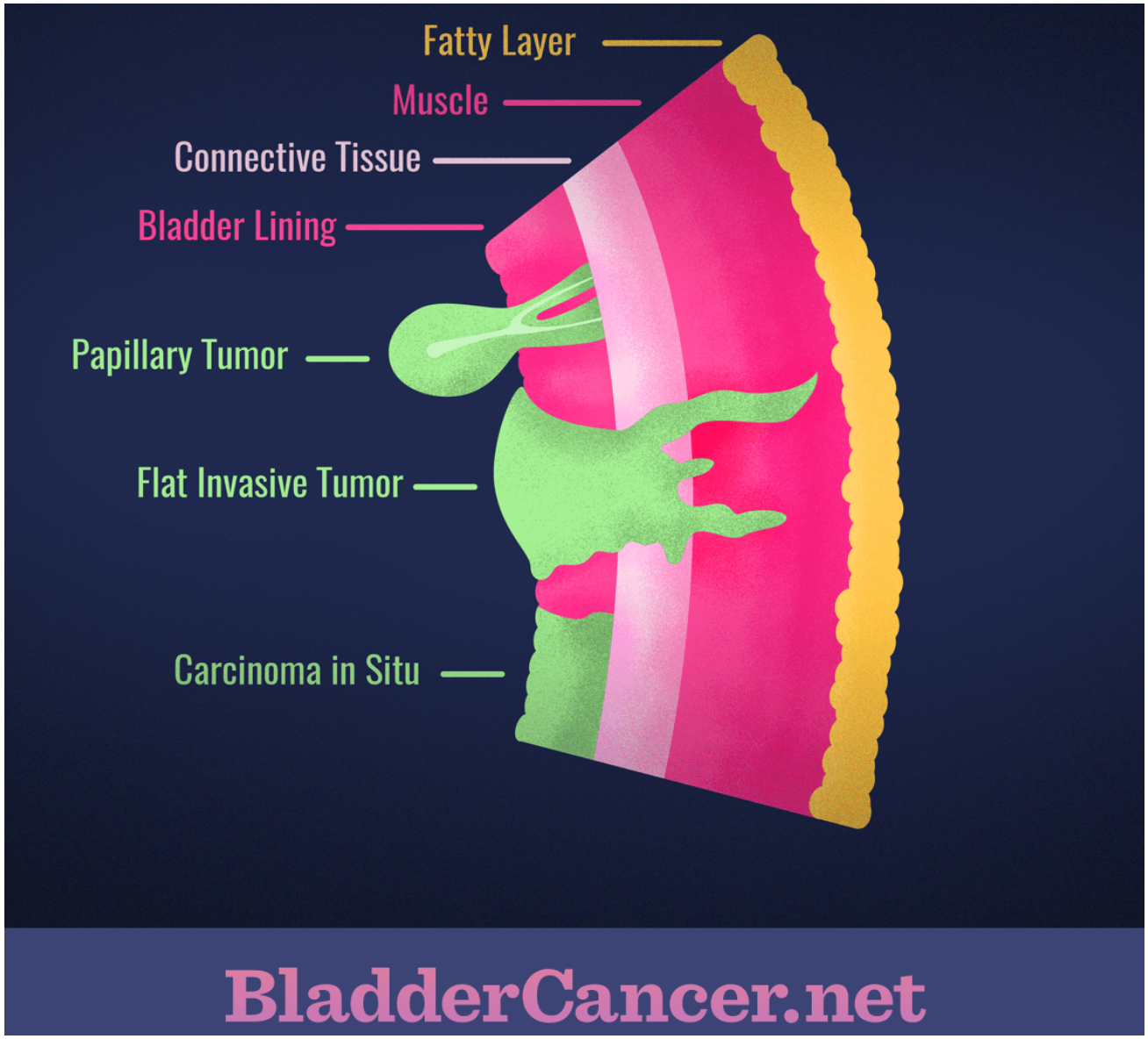

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
C659, C669, C670-C679, C680-C689
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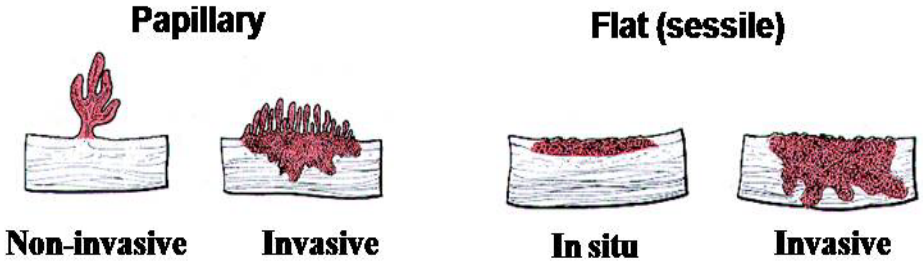
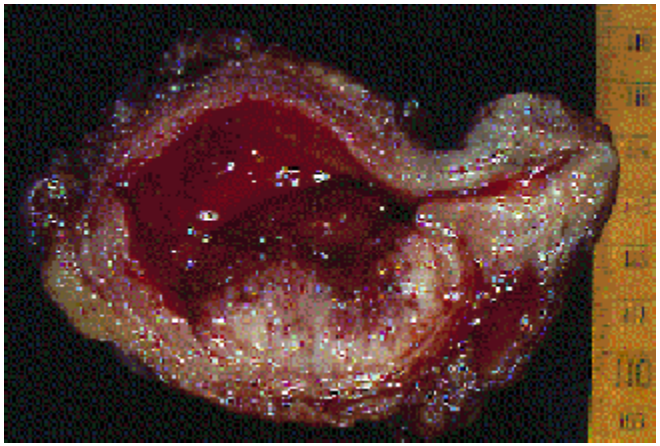


Bladder Tumor



Courtesy Jean-Loup Huret reprinted from Atlas Genet Cytogenet Oncol Haematol. October 2003. van Tilborg A

A, van Rhijn BW. Bladder: Urothelial carcinomas. URL: http://atlasgeneticsoncology.org/Tumors/bladID5001.html, by permission of the Atlas.

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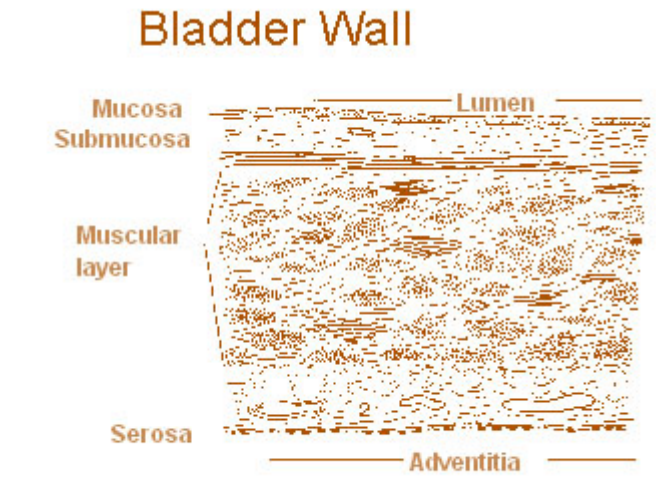
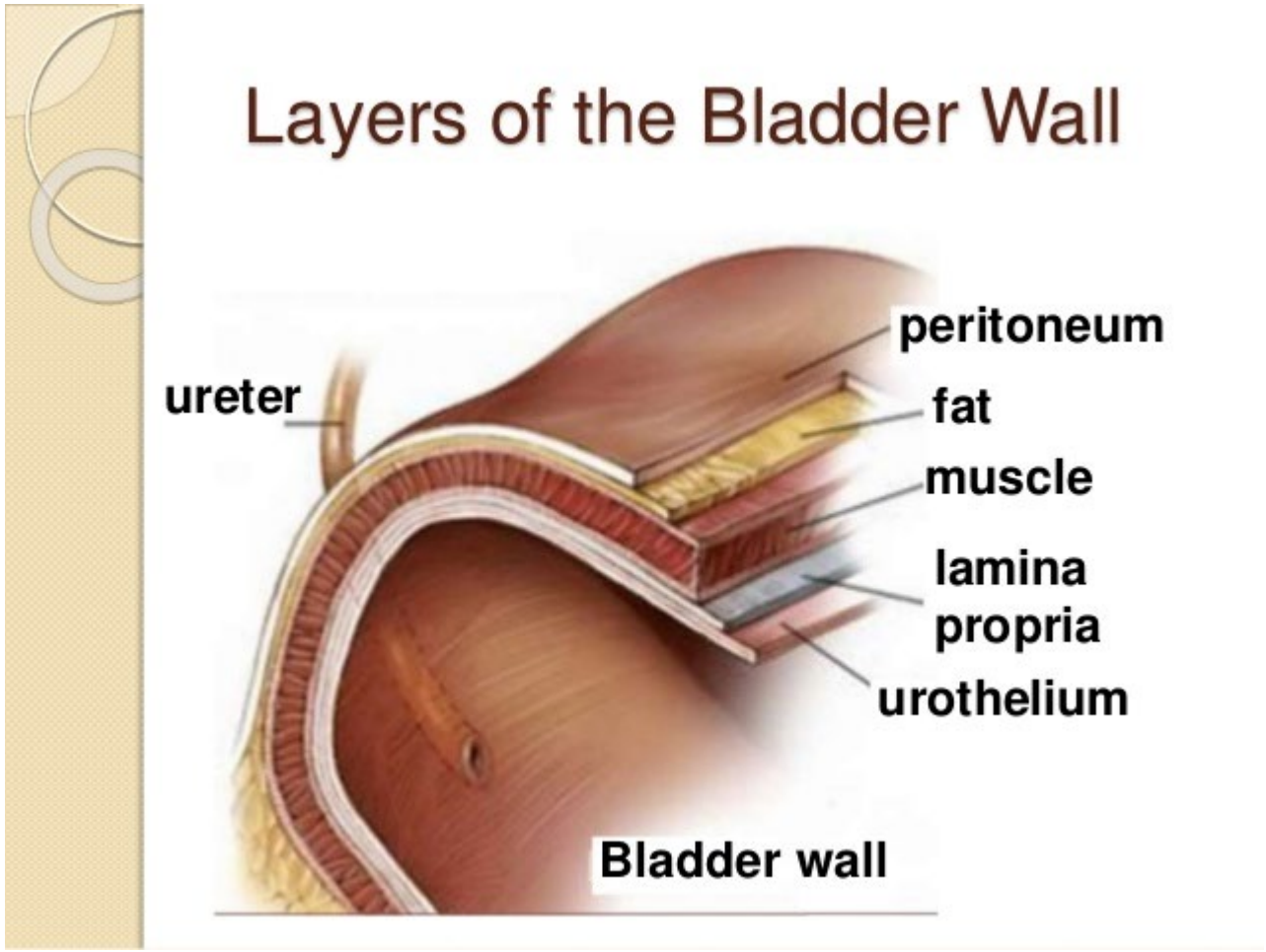
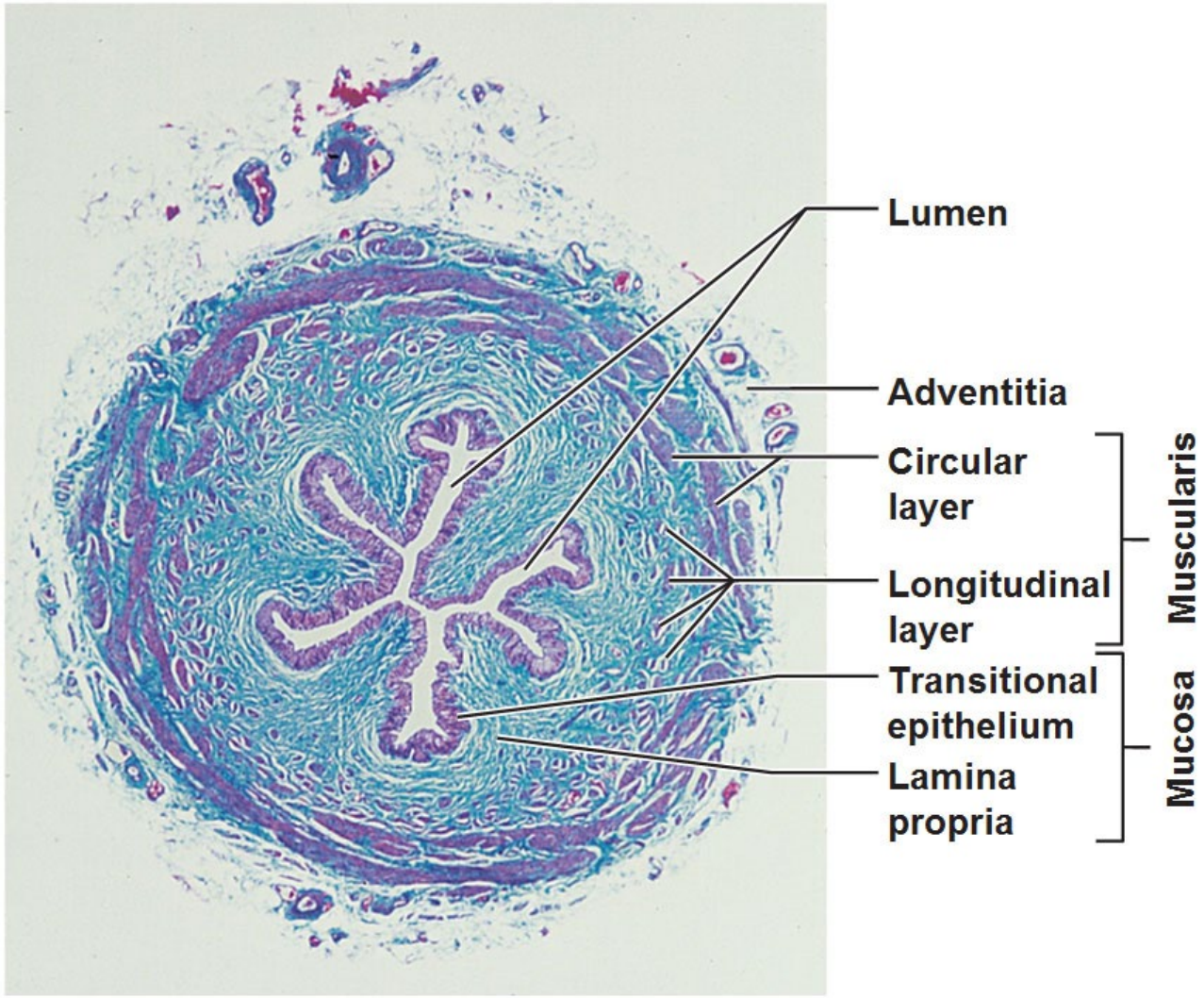


Source: Feneis, Pocket Atlas of Human Anatomy, 2nd ed.


Layers of the Bladder Wall
peritoneum
ureter
fat
muscle
lamina
propria
urothelium
Bladder wall

Microscopic Structure of the Ureter



Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Bones
- Brain
- Regional and distant lymph nodes for the primary site being abstracted as identified in Summary Staging Manual
- Involvement of the pelvic or abdominal wall
- Liver
- Lung

Note 2: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include:

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors.

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

Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Single Tumor

Rule M2 Abstract a **single primary**[i] when there is a **single tumor**.

Note 1: A single tumor is always a single primary.
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor.

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

Multiple Tumors

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

- In the urinary system (see Table 1) **AND** in a site other than the urinary system
  Example: Patient has urothelial carcinoma of the bladder and non-metastatic adenocarcinoma of the lung. The lung is not a urinary site. Abstract two primaries.
- Non-synchronous tumors other than urothelial carcinoma and urothelial carcinoma subtypes in multiple urinary sites (see Rule M14)

Rule M3 Abstract **multiple primaries**[ii] when there are:

- Separate/non-contiguous tumors in both the right **AND** left **renal pelvis AND**
- No other urinary sites are involved with separate/non-contiguous tumors

Note 1: Only abstract a single primary when pathology confirms tumor(s) in the contralateral renal pelvis are metastatic.
Note 2: This rule is used **only** when there is **no involvement** by separate/non-contiguous tumors in the ureter(s), bladder, or urethra.

Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M4 Abstract **multiple primaries**[ii] when there are:

- Separate/non-contiguous tumors in the right **AND** left **ureter AND**
- No other urinary sites are involved with separate/non-contiguous tumors

Note 1: Only abstract a single primary when pathology confirms tumor(s) in contralateral ureter are metastatic.

Note 2: This rule is used **only** when there is **no involvement** by separate/non-contiguous tumors in the renal pelvis, bladder, and urethra.

Rule M5 Abstract a **single primary**[i] when **synchronous** tumors are **noninvasive in situ /2** urothelial carcinoma (flat tumor) **8120/2 in the following sites:**

- **Bladder C67_ AND**
- One or both **ureter**(s) **C669**

Note 1: No other urinary organs are involved.

Note 2: Use this rule **ONLY** for noninvasive in situ urothelial carcinoma (may be called noninvasive urothelial carcinoma or noninvasive flat tumor). For other histologies, continue through the rules.

Note 3: Urothelial carcinoma in situ spreads by intramucosal extension and may involve large areas of mucosal surface. The default for these cases is coding a bladder primary.

Rule M6 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor.

Note 1: Abstract both the invasive and in situ tumors.

Note 2: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression.

Note 3: This rule is based on long-term epidemiologic studies of recurrence intervals. The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were also authors, co-authors, or editors of the AJCC Staging

Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M7 Abstract a **single primary**[i] when the patient has multiple occurrences of /2 urothelial carcinoma in the **bladder**. Tumors may be any combination of:

- In situ urothelial carcinoma **8120/2 AND/OR**
- Papillary urothelial carcinoma noninvasive **8130/2** (does **not** include micropapillary subtype)

Note 1: Timing is irrelevant. Tumors may be synchronous or non-synchronous.

Note 2: Abstract only one /2 urothelial bladder primary per the patient's lifetime.

Note 3: There are no /2 subtypes for urothelial carcinoma with the exception of papillary urothelial carcinoma.

Example: On 1/3/2018, the patient had a TURB with a diagnosis of in situ urothelial carcinoma 8120/2. On 5/8/2019, pathology from TURB is papillary urothelial carcinoma non-invasive 8130/2. This is a single primary; the papillary urothelial carcinoma is recorded as a recurrence for those registrars who collect recurrence data.

Rule M8 Abstract **multiple primaries**[ii] when the patient has **micropapillary** urothelial carcinoma **8131/3 of the bladder AND** a urothelial carcinoma **8120/3** (including papillary **8130/3**) **of the bladder**.

Note 1: This is a new rule for 2018.

Note 2: Micropapillary urothelial cell carcinoma is an extremely aggressive neoplasm. It is important to abstract a new primary to capture the incidence of micropapillary urothelial carcinoma. Micropapillary is excluded from the typical "NOS and subtype/variant" rule (same row in Table 2).

Rule M9 Abstract a **single primary**[i] when the patient has multiple **invasive** urothelial cell carcinomas in the **bladder**. All tumors are **either**:

- Multiple occurrences of urothelial or urothelial subtypes (with exception of micropapillary) **OR**
- Multiple occurrences of micropapillary

Note 1: Timing is irrelevant. Tumors may be synchronous or non-synchronous.

Note 2: Abstract only one /3 invasive urothelial bladder primary **AND** only one micropapillary urothelial 8131/3 bladder primary per the patient's lifetime.

- An occurrence of micropapillary and an occurrence of urothelial carcinoma would be multiple primaries (see previous rules).

Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M10 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being **clinically disease-free** for **greater than three years** after the original diagnosis or last recurrence.

Note 1: This rule **does not apply** when both/all tumors are urothelial carcinoma of the bladder (all subtypes/variants of 8120 except for 8131).

Note 2: **Clinically** disease-free means that there was **no evidence** of recurrence on follow-up.

- Scans are NED
- Urine cytology is NED
- Scopes are NED

Note 3: When there is a recurrence within three years of diagnosis, the "**clock**" starts over. The time interval is calculated from the **date of last recurrence**.

Note 4: When it is **unknown/not documented** whether the patient had a recurrence, default to **date of diagnosis** to compute the time interval.

Note 5: The physician may state this is a **recurrence,** meaning the patient had a previous urinary site tumor and now has another urinary site tumor. **Follow the rules**; do not attempt to interpret the physician's statement.

Example: Patient is diagnosed with multifocal/multicentric urothelial carcinomas in the ureter and renal pelvis in January 2018. Both the kidney and ureter are surgically removed. In June 2022 the patient presents with tumor in the contralateral ureter. The physician states this is a recurrence of the original urothelial carcinoma. Code a new primary for the 2022 ureter carcinoma.

Rule M11 Abstract a **single primary**[i] when there are **urothelial carcinomas** in multiple urinary organs.

Note 1: This rule is **ONLY** for urothelial carcinoma **8120** and all subtypes/variants of urothelial carcinoma. This rule does not apply to any other carcinomas or sarcomas.

Note 2: Behavior is irrelevant.

Note 3: This rule applies to multifocal/multicentric carcinoma which involves two or more of the following urinary sites:

- Renal pelvis
- Ureter
- Bladder
- Urethra

Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M12 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3 of **Table 2** in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS**: Leiomyosarcoma 8890/3 and liposarcoma 8850/3 are both subtypes of sarcoma NOS 8800/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Verrucous carcinoma 8051 is a subtype of squamous cell carcinoma NOS 8070; giant cell urothelial carcinoma 8031 is a subtype of urothelial carcinoma 8120. They are distinctly different histologies. Abstract multiple primaries.

Rule M13 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are on **different rows** in **Table 2** in the Equivalent Terms and Definitions. Timing is irrelevant.

Note: Each row in the table is a **distinctly different** histology.

Example: Small cell neuroendocrine carcinoma 8041 and urothelial carcinoma 8120 are on different rows of Table 2. Abstract two primaries, one for the small cell neuroendocrine carcinoma and a second for the urothelial carcinoma.

Rule M14 Abstract **multiple primaries**[ii] when the ICD-O **site** code differs at the **second** (C**X**xx) and/or **third** (Cx**X**x) character.

Rule M15 Abstract a **single primary**[i] when **synchronous**, separate/non-contiguous tumors are on the **same row** in **Table 2** in the Equivalent Terms and Definitions.

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3) **OR**
- A NOS histology in column 3 with an indented subtype/variant

Example: TURBT shows invasive papillary urothelial carcinoma 8130/3 and CIS/in situ urothelial carcinoma 8120/2. Abstract a single primary. Papillary urothelial carcinoma and urothelial carcinoma are on the same row in Table 3.

Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule M16 Abstract a **single primary**[i] (the invasive) when an **in situ** tumor is diagnosed **after** an **invasive** tumor **AND** tumors occur in the **same** urinary site.

Note 1: The rules are hierarchical. Only use this rule when previous rules do not apply.

Note 2: The tumors may be a NOS and a subtype/variant of that NOS. See **Table 2** in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

Note 3: Once the patient has an invasive tumor, the subsequent **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.

Rule M17 Abstract a **single primary**[i] (the invasive) when an invasive tumor is diagnosed **less than or equal to 60** days **after** an **in situ** tumor **AND** tumors occur in the **same** urinary site.

Note 1: The rules are hierarchical. Only use this rule if none of the previous rules apply.

Note 2: The tumors may be an NOS and a subtype/variant of that NOS.

Note 3: When the case has been abstracted, change behavior code on original abstract from /2 to /3. Do not change date of diagnosis.

Note 4: If the case has already been submitted to the central registry, report all changes.

Note 5: The physician may stage both tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

Note 6: See the **COC** and **SEER** manuals for **instructions** on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M18 Abstract a **single primary**[i] when tumors do not meet any of the above criteria.

Note: Use this rule as a last resort. Please confirm that you have not overlooked an applicable rule.

This is the end of instructions for Multiple Tumors.

[i] Prepare one abstract. Use the histology coding rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

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

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

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

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

Code the **most specific** pathology/tissue from either **resection** or **biopsy**.

Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

Use documentation in the following priority order to identify the histology type(s):

1. **Tissue or pathology report from primary site** (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol

   Note 1: Addendums and comments on the pathology report are given a high priority because they often contain information about molecular testing, genetic testing, and/or special stains which give a more specific diagnosis.
   Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority. The final diagnosis is often the synoptic CAP report.

Note 3: The CAP protocol is a checklist which:

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

2. **Cytology** (usually urine)
3. Tissue/pathology from a metastatic site
   Note 1: Code the behavior /3.
   Note 2: The **tissue** from a **metastatic** site often shows **variations** from the primary tumor. When it is the only tissue available, it is **more accurate** than a **scan and only physician documentation**.
4. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following **priority order:**
   A. Treatment Plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that **refers to original pathology, cytology, or scan(s)**
   D. Physician's **reference to** type of cancer (**histology**) in the medical record
   Note 1: Code the specific histology when documented.
   Note 2: Code the histology to 8000 (cancer/malignant neoplasm NOS) or as stated by the physician when nothing more specific is documented.
5. **Scans: CT, MRI.** There is **no priority** order because scans are not a very reliable method for **identifying** specific **histology**(ies) for these sites.

Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Note: Only code **differentiation** or **features** when there is a **specific code** for the NOS with differentiation or the NOS with features in **Table 2** or the **ICD-O and all updates. This instruction applies to single and multiple histologies.**

Coding Histology

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

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

   Example 1: Diagnosis for a single tumor is adenocarcinoma 8140 with the majority or predominant part of tumor being endometrioid carcinoma 8380. Code the subtype/variant: endometrioid carcinoma 8380.
   Example 2: Diagnosis for a single tumor is small cell neuroendocrine carcinoma 8041 with minority of tumor being large cell neuroendocrine tumor 8013. Code the subtype/variant: large cell neuroendocrine tumor 8013.
   Example 3: Diagnosis for a single tumor is sarcoma NOS 8800/3 with a component of leiomyosarcoma 8890/3. Code the subtype/variant: leiomyosarcoma 8890/3.

   Note 1: The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.
   Example: When the diagnosis is adenocarcinoma with a clear cell **carcinoma** component, code clear cell carcinoma 8310.
   Negative Example: When the diagnosis is simply adenocarcinoma with a clear cell component, code adenocarcinoma NOS 8140. Do not assume this is a clear cell carcinoma. This could be clear cell differentiation or features.

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

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

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

      Example: Outpatient biopsy says probably papillary urothelial cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology papillary urothelial cell carcinoma. The case meets the criteria in #**3A.**

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

      Example 1: The pathology diagnosis is sarcoma NOS consistent with leiomyosarcoma. The oncology consult says the patient has leiomyosarcoma of the bladder. This is clinical confirmation of the diagnosis, code leiomyosarcoma. The case meets the criteria in **bullet 1**.

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

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

List of Ambiguous Terminology

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

4. **DO NOT CODE** histology when described as:

- Architecture
- Foci; focus; focal
- Growth pattern
- Pattern

Single Tumor

Rule H1 Code the histology when only **one histology** is present.

Note 1: Use **Table 2** to code histology. New codes, terms, and synonyms are included in **Table 2** and coding errors may occur if the table is not used.

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

Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 2, ICD-O or all updates.

Note 4: Only code squamous cell carcinoma (8070) when there are no other histologies present (pure squamous cell carcinoma).

Note 5: Only code adenocarcinoma (8140) when there are no other histologies present (pure adenocarcinoma).

Rule H2 Code the **invasive** histology **when in situ** and **invasive** histologies are present in the **same tumo**r.

Rule H3 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Papillary urothelial carcinoma **8130** and a subtype/variant of papillary urothelial carcinoma
- Rhabdomyosarcoma **8900** and a subtype/variant of rhabdomyosarcoma
- Sarcoma **8800** and a subtype/variant of sarcoma
- Small cell neuroendocrine carcinoma **8041** and a subtype/variant of small cell neuroendocrine carcinoma
- Squamous cell carcinoma **8070** and a subtype/variant of squamous cell carcinoma
- Urothelial carcinoma **8120** and a subtype/variant of urothelial carcinoma

Note: Use **Table 2** to identify NOS histologies and subtypes/variants.

Rule H4 Code mixed small cell carcinoma **8045** when the final diagnosis is any of the following:

- Small cell neuroendocrine mixed with any other type of **carcinoma** (does not apply to sarcoma)
- Two or more subtypes/variants of small cell neuroendocrine carcinoma
- Subtype/variant of small cell neuroendocrine mixed with any other **carcinoma** (does not apply to sarcoma)

Example: Diagnosis from TURB is urothelial carcinoma **and** small cell neuroendocrine carcinoma. Code mixed small cell carcinoma 8045.

Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Rule H5 Code mixed urothelial carcinoma as follows:

- Code **8120** when urothelial is mixed with:
  - Adenocarcinoma or adenocarcinoma subtypes
  - Squamous cell carcinoma or squamous cell carcinoma subtypes
- Code **8130** when papillary urothelial is mixed with:
  - Adenocarcinoma or adenocarcinoma subtypes
  - Squamous cell carcinoma or squamous cell carcinoma subtypes
- Code **8131/3** when micropapillary urothelial is mixed with:
  - Adenocarcinoma or adenocarcinoma subtypes
  - Squamous cell carcinoma or squamous cell carcinoma subtypes

Note: Adenocarcinoma and subtypes/variants as well as squamous cell carcinoma and subtypes/variants are coded ONLY when pure (not mixed with any other histology).

Example: Pathology says majority of tumor is squamous cell carcinoma 8070/3 with a minority composed of papillary urothelial cell carcinoma 8130/3. Code the papillary urothelial cell carcinoma 8130/3. The squamous cell carcinoma is not pure and cannot be coded.

This is the end of instructions for Single Tumor.

Code the histology using the rule that fits the case.

Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9993 and Kaposi sarcoma M9140)

Multiple Tumors Abstracted as a Single Primary

Rule H6 Code the histology when only **one** histology is present in **all** tumors.

Note 1: Use **Table 2** to code histology. New codes, terms, and synonyms are included in **Table 2** and coding errors may occur if the table is not used.

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

Note 3: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 2, ICD-O or all updates.

Note 4: Only code squamous cell carcinoma (8070) when there are no other histologies present (pure squamous cell carcinoma).

Note 5: Only code adenocarcinoma (8140) when there are no other histologies present (pure adenocarcinoma).

Rule H7 Code the **invasive** histology when there are invasive and in situ histologies:

- Mixed in each of the tumors **OR**
- In separate tumors (one or more invasive and one or more in situ)

Rule H8 Code the **subtype/variant** when **all multifocal/multicentric** tumors are a **NOS** and **a single subtype/variant** of that NOS such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Papillary urothelial carcinoma **8130** and a subtype/variant of papillary urothelial carcinoma
- Rhabdomyosarcoma **8900** and a subtype/variant of rhabdomyosarcoma
- Sarcoma **8800** and a subtype/variant of sarcoma
- Small cell neuroendocrine carcinoma **8041** and a subtype/variant of small cell neuroendocrine carcinoma
- Squamous cell carcinoma **8070** and a subtype/variant of squamous cell carcinoma
- Urothelial carcinoma **8120** and a subtype/variant of urothelial carcinoma

Note 1: Use **Table 2** to identify NOS histologies and subtypes/variants.

Note 2: All tumors may be mixed histologies (NOS and a subtype/variant of that NOS) **OR** one tumor may be a NOS histology and the other tumor a subtype/variant of that NOS.

Rule H9 Code mixed small cell carcinoma **8045** when the final diagnosis **for all tumors** is any of the following:

- Small cell neuroendocrine mixed with any other type of **carcinoma** (does not apply to sarcoma)
- Two or more subtypes/variants of small cell neuroendocrine carcinoma
- Subtype/variant of small cell neuroendocrine mixed with any other carcinoma (does not apply to sarcoma)

Example: Diagnosis from TURB is urothelial carcinoma **and** small cell neuroendocrine carcinoma. Code mixed small cell carcinoma 8045.

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

Code the histology using the rule that fits the case.

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Introduction

Note 1: Tables and rules refer to ICD-O rather than ICD-O-3. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

***Note 2*:** 2007 MPH Rules and 2021 Solid Tumor Rules are used based on date of diagnosis

- Tumors diagnosed 01/01/2007 through 12/31/2020: Use 2007 MPH Rules and 2007 General Instructions
- Tumors diagnosed 01/01/2021 and later: Use 2021 Solid Tumor Rules and Solid Tumor General Instructions
- The original tumor diagnosed ***before*** 01/01/2021 and a subsequent tumor diagnosed 01/01/2021 or later in the same primary site: Use the 2021 Solid Tumor Rules and Solid Tumor General Instructions

Note 3*:** Melanoma can also start in the mucous membranes of the mouth, anus and vagina, in the eye or other places in the body where melanocytes are found. This scheme is used ***only for melanomas that ***occur on the skin***.

Note 4: The WHO Classification of Skin Tumors 4th Ed does not include ICD-O codes for tumors with mixed melanoma subtypes/variants

Note 5: Cutaneous melanoma starts in the melanocytes of the skin. Melanocytes lie in the epidermis, the outermost layer of the skin. Melanocytes often cluster together and form moles (nevi). Most moles are benign, but some may become malignant melanomas. Melanomas are divided into 5 main types, depending on their location, shape, and whether they grow outward or downward into the dermis:

- Acral melanoma: occurs on the palms of the hand, soles of the feet, or nail beds
- Desmoplastic melanoma: is a rare malignant melanoma marked by non-pigmented lesions on sun exposed areas of the body
- Lentigo maligna: usually occur on the faces of elderly people
- Superficial spreading or flat melanoma: grows outwards at first to form an irregular pattern on the skin with an uneven color
- Nodular melanoma: are lumpy and often blue-black in color and may grow faster and spread downwards

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Changes from 2007 MPH Rules

These changes are effective with cases diagnosed 1/1/2021 and later. WHO 4th Ed Classification of Skin Tumors was published in 2018.

1. 2007 Rules instruct "Code the histology from the most representative specimen." For all sites except breast and CNS, the 2021 Rules instruct "Code the most specific histology from biopsy or resections". When there is a discrepancy between the biopsy and resection (two distinctly different histologies), code the histology from the most representative specimen (the greater amount of tumor)." **This instruction applies to the 2021 cutaneous melanoma solid tumor rules**.
2. Early/evolving melanoma in situ (8720/2) and early/evolving melanoma invasive (8720/3) are reportable for cases diagnosed 1/1/2021 and later. Please refer to SEER Program Coding and Staging Manual 2021 for additional information on reportable neoplasms.
3. New histology **terms** are included (identified by asterisks (*) in the histology table in the Terms and Definitions). No new cutaneous melanoma ICD-O histology **codes** have been proposed by WHO.
4. Some histologies are rare and may not be listed in the tables; refer to ICD-O and all updates. If the histology is not found in the tables or ICD-O, submit a question to Ask a SEER Registrar.
5. WHO 4th Ed Skin Tumors now classifies melanocytic tumors into two groups:
   A. Melanomas arising in sun-exposed skin
   B. Melanomas arising at sun-shielded sites or without known etiological association with UV radiation exposure

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
  Note: "And" and "with" are used as synonyms when describing multiple histologies within a single tumor.
- Giant pigmented nevus; giant congenital nevus
- Mixed epithelioid and spindle cell melanoma (8770); Epithelioid melanoma and spindle cell melanoma
- Melanoma in situ, superficial spreading type; low-cumulative sun damage (CSD) melanoma in situ
- Mole; Nevus
- Simultaneous; existing at the same time; concurrent
- Site; topography
- Superficial spreading melanoma; low-cumulative sun damage (CSD) melanoma
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician's statement that the term is malignant/melanoma**
  - These terms are used **ONLY** to determine multiple primaries
  - **Do not** use these terms for casefinding or determining reportability
- Type; subtype; variant

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terms that are not Equivalent or Equal

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

- Component is not equivalent to subtype/type/variant
  Note 1: Component is only coded when the pathologist specifies the component as a second ***melanoma***
  Note 2: Examples provided in H rules Coding Histology section
- Phenotype is not equivalent to subtype/type/variant

Synonyms for In Situ

- Behavior code 2
- Clark level 1 (limited to the epithelium)
- Hutchinson freckle (See Synonyms for Hutchinson Freckle)
- Intraepidermal, NOS
- Intraepithelial, NOS
- Lentigo maligna
- Noninvasive
- Precancerous melanoma of Dubreuilh
- Precancerous melanosis (C44_)
- Stage 0
- Tis

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
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Synonyms for Hutchinson Freckle

- Circumscribed precancerous melanosis
- Intraepidermal malignant melanoma
- Lentigo maligna
- Precancerous melanosis of Dubreuilh

Anatomical Dermatology Terms

Term	Definition
Cutaneous	Pertaining to skin
Dermal	Pertaining to skin
Epidermal	Pertaining to upon the skin
Hypodermic	Pertaining to below the skin
Intradermal	Pertaining to within the skin
Subcutaneous	Pertaining to under the skin
Ungual	Pertaining to the nail

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Table 1: Primary Sites and Laterality

Table 1 contains terms used in **clinical diagnosis,** and less frequently the **operative and pathology reports** to describe the **location** of the skin lesion. Find the **term** in Column 1 and use the **site code** in Column 2. Column 3 notes whether the site requires **laterality** to be coded.

Note: Excludes melanoma of non-skin sites (excludes any sites other than C44_)

Terminology	Site Term and Code	Laterality Required
Skin of lip, NOS Skin of lower lip Skin of upper lip	Skin of lip, NOS **C440**	**No**
Eyelid Lid, NOS Palpebra Horizontal palpebra fissure Canthus Inner canthus Lateral canthus Lower lid Medial canthus Meibomian gland Outer canthus Pretarsal space Supratarsel crease Upper lid	Eyelid **C441**	**Yes**

Cutaneous Melanoma Equivalent Terms and Definitions
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Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
External ear Auricle, NOS Pinna Ceruminal gland Concha Ear, NOS Ear lobule Earlobe External auditory canal Auditory canal, NOS Auricular canal, NOS External auricular canal Ear canal External auditory meatus Helix Skin of auricle Skin of ear, NOS Tragus	External ear **C442**	**Yes**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of other & unspecified parts of face Skin of: Cheek Chin Face Forehead Jaw Nose Temple Ala nasi Chin, NOS Columnella Eyebrow Brow External cheek External nose Forehead, NOS Lid-cheek junction Nasaljugal groove Temple, NOS	Skin of other and unspecified parts of face **C443**	**Yes**
Skin of scalp and neck Skin of head, NOS Skin of neck Skin of scalp Scalp, NOS Skin of cervical region	Skin of scalp and neck **C444**	**Yes (New)**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of trunk Skin of: Abdomen Abdominal wall Anus Axilla Back Breast Buttock Chest Chest wall Flank Groin Perineum Thoracic wall Thorax Trunk Umbilicus Gluteal region Infraclavicular region Inguinal region Sacrococcygeal region Scapular region Perianal skin Umbilicus, NOS	Skin of trunk **C445**	**Yes**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of upper limb and shoulder Skin of: Antecubital space Arm Elbow Finger Forearm Hand Palm Shoulder Thumb Upper limb Wrist Fingernail Nail Bed Palmar skin	Skin of upper limb and shoulder **C446**	**Yes**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of lower limb and hip Skin of: Ankle Calf Foot Heel Hip Knee Leg Lower limb Popliteal space Thigh Toe Plantar skin Sole of foot Toenail	Skin of lower limb and hip **C447**	**Yes**
Overlapping lesion of skin	Overlapping lesion of skin **C448** For Head and Neck: Do not use C448 for overlapping lesions of the Head & Neck. Assign the primary site code for the site where the bulk of the tumor is or where the epicenter is; do not use code C448.	**No**
Skin, NOS ***Note:*** Code to Skin, NOS (C44.9) if a patient is diagnosed with metastatic melanoma and the primary site is not identified.	Skin, NOS **C449**	**No**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Table 2: Specific Histologies, NOS, and Subtype/Variants

Use Table 2 as directed by the **Histology Rules** to assign the more common histology codes for melanotic skin tumors

Note 1: Rare histologies may not be listed in the table. When a histology term is not found, reference ICD-O and all updates.
Note 2: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 2, ICD-O or ICD-O updates.
Note 3: Behavior codes are listed when the term has only one possible behavior (either a /2 or /3). For histologies which may be either /2 or /3, a behavior code is not listed. Code behavior as documented in the pathology report.
Note 4: Only use the histology code from the table when the diagnosis is **EXACTLY** the term listed.
Note 5: **New histology terms are identified by asterisk (*)**

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

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

Table begins on the next page

***New terms approved by IARC/WHO Committee for ICD-O**
****Terms approved by standard setters and are not listed in WHO or ICD-O**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

NOS Histology Terms and Codes	Synonyms	Subtypes/Variants
Melanoma, NOS **8720** ***Note***: Sarcomatoid melanoma is a rare subtype of melanoma characterized by almost complete loss of melanocytic differentiation both morphologically and phenotypically, with the bulk of the tumor being replaced by a spindle cell, sarcomatoid component. Use code 8772/3, spindle cell melanoma.	Melanoma in situ **8720/2** Early/Evolving melanoma in situ** **8720/2** Nevoid melanoma **8720/3** Early/Evolving invasive melanoma** **8720/3**	Acral melanoma*/acral lentiginous melanoma, malignant **8744/3** Amelanotic melanoma **8730/3** Balloon cell melanoma **8722/3** Desmoplastic melanoma/desmoplastic melanoma, amelanotic/neurotropic melanoma, malignant **8745/3*** Epithelioid cell melanoma **8771/3** Lentigo maligna/Hutchinson melanotic freckle **8742/2 /** Lentigo maligna melanoma/Melanoma in Hutchinson melanotic freckle **8742/3** Low cumulative sun damage melanoma*/superficial spreading melanoma **8743/3** Melanoma arising in a blue nevus **8780/3*** Malignant melanoma arising in giant congenital nevus*/malignant melanoma in giant pigmented nevus **8761/3** Malignant melanoma in a precancerous melanosis **8741/3** Malignant melanoma, regressing **8723/3** Malignant Spitz tumor*/mixed epithelioid and spindle cell melanoma **8770/3** Nodular melanoma **8721/3** Spindle cell melanoma **8772/3** Spindle cell melanoma, type A **8773/3** Spindle cell melanoma, type B **8774/3**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Table 3: Non-Reportable Neoplasms

Table 3 lists non-reportable terms and codes used in the diagnosis of cutaneous melanotic neoplasms. ***This table is intended to be a reference only and may not be complete***. Please refer to your standard setter program manual for additional information on reportable neoplasms.

Non-Reportable Histology Term	Non-Reportable Histology Code
Pigmented nevus, NOS Nevus, NOS Melanocytic nevus Hairy nevus Nevus spilus Meyerson nevus Deep penetrating nevus Combined nevus Genital nevus Conjunctival nevus	**8720/0**
Balloon cell nevus	**8722/0**
Halo nevus Regressing nevus	**8723/0**
Neuronevus	**8725/0**
Magnocellular nevus Melanocytoma, NOS	**8726/0**
Dysplastic nevus	**8727/0**
Nonpigmented nevus Achromic nevus	**8730/0**
Junctional nevus, NOS Intraepidermal nevus Junction nevus	**8740/0**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

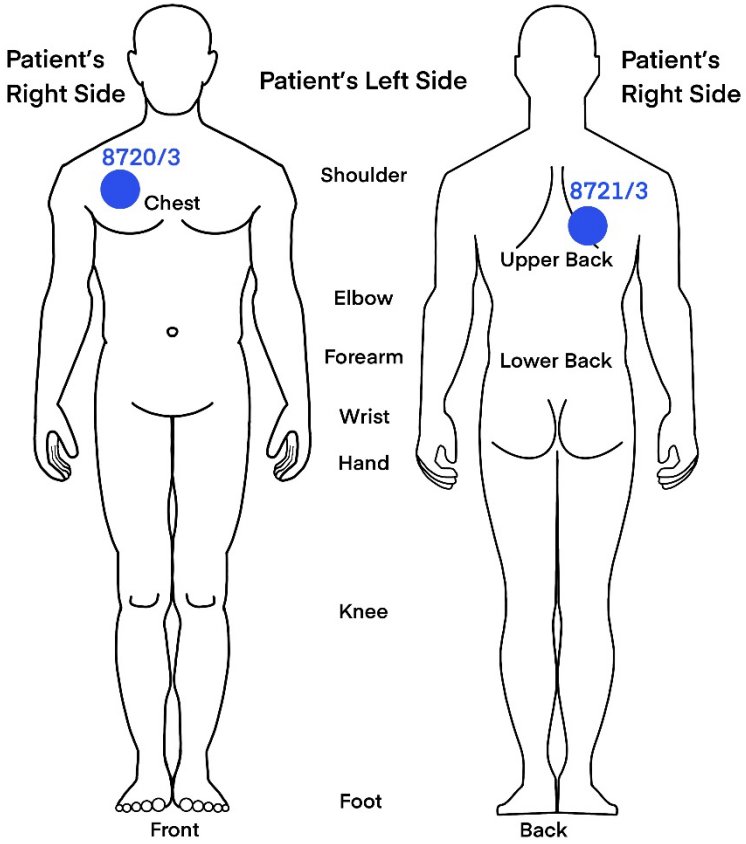
Non-Reportable Histology Term	Non-Reportable Histology Code
Lentiginous melanocytic nevus Simple lentigo Lentigo simplex	**8742/0**
Acral nevus	**8744/0**
Dermal nevus Intradermal nevus Stromal nevus	**8750/0**
Compound nevus Dermal and epidermal nevus	**8760/0**
Congenital melanocytic nevus, NOS Giant pigmented nevus, NOS Intermediate and giant congenital nevus	**8761/0** **8761/0** **8761/1**
Proliferative dermal lesion in congenital nevus Proliferative nodule in congenital melanocytic nevus	**8762/1**
Epithelioid and spindle cell nevus Juvenile melanoma Juvenile nevus Spitz nevus Spitz nevus, atypical Pigmented spindle cell nevus of Reed Pigmented spindle cell Spitz nevus	**8770/0**
Epithelioid cell nevus	**8771/0**
Spindle cell nevus, NOS	**8772/0**
Blue nevus, NOS Jadassohn blue nevus Pigmented epithelioid melanocytoma Blue nevus, epithelioid	**8780/0** **8780/0** **8780/1** **8780/1**
Cellular blue nevus	**8790/0**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Non-Reportable Histology Term	Non-Reportable Histology Code
Intermediate lesion Melanocytic neoplasm of low malignant potential Melanocytic tumor of uncertain malignant potential (MELTUMP) Superficial atypical melanocytic proliferation of uncertain significance (SAMPUS) Primary acquired melanosis	No ICD-O code

Illustrations

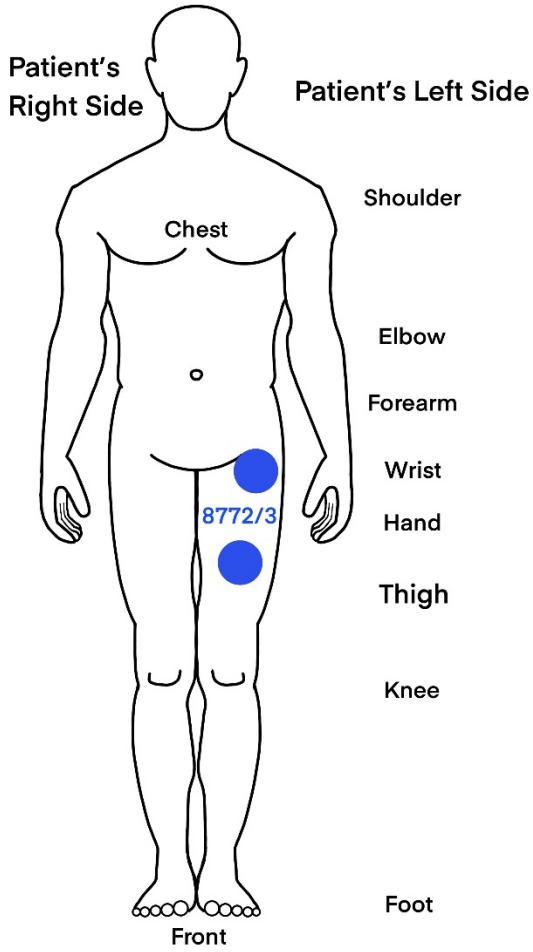
Explanatory illustrations for Multiple Primary Rule M6



Example 1: Both lesions are located on the right side of the body and sites are chest C445 and back C445. Histology for the chest lesion is melanoma, NOS (8720/3) and the back lesion is nodular melanoma (8721/3). Abstract a single primary.

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Explanatory illustrations for Multiple Primary Rule M6



Example 2: Both lesions are located on the left leg. One lesion is spindle cell melanoma (8772/3) located on the front of the left hip C447. The other lesion is spindle cell melanoma (8772/3) located on the front of the left thigh C447.

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

- Distant metastasis in skin, subcutaneous tissue including muscle
- Bone
- Brain
- Regional lymph nodes as identified in Summary Stage Manual
- Distant lymph nodes as identified in Summary Stage Manual
- Liver
- Lung
- In-transit metastases – metastases which occur along the lymph pathways between the primary tumor > 2 cm from the scar and the regional lymph nodes
- Satellites – new tumor within a radius of 2 cm from the scar after removal of primary tumor. Satellites may be caused by remains of the primary tumor.

Note 2: 2007 MPH Rules and 2021 Cutaneous Melanoma Solid Tumor Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2020: Use 2007 MPH Rules and 2007 General Instructions
- Tumors diagnosed 01/01/2021 and later: Use 2021 Solid Tumor Rules
- The original tumor diagnosed ***before*** 1/1/2021 and a subsequent tumor diagnosed 1/1/2021 or later in the same primary site: Use the 2021 Solid Tumor Rules

Unknown if Single or Multiple Melanomas

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there is a **single** melanoma or **multiple** melanomas.

Note 1: Use this rule only after all information sources have been exhausted.

Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Melanoma

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

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Single Melanoma

Note 1: Do not use the rules for melanoma described as metastasis
Note 2: Includes combinations of in situ and invasive

Rule M2 Abstract a **single primary**[i] when there is a **single melanoma.**
Note 1: A single melanoma is always a single primary
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.
Note 3: The tumor may have in situ and invasive components.
Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Melanoma.

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

Multiple Melanomas

Note 1: Multiple melanomas may be a single primary or multiple primaries
Note 2: Do not use the rules for melanoma described as metastasis
Note 3: Includes combinations of in situ and invasive

Rule M3 Abstract **multiple primaries**[ii] when there are separate, non-contiguous melanomas in sites with ICD-O site codes that **differ** at the second (C**X**xx), third (Cx**X**x) or fourth (C44**X**) character.
Note: This applies to a melanoma of unknown primary and a known cutaneous melanoma primary

Rule M4 Abstract **multiple primaries**[ii] when there are separate, non-contiguous melanomas with **different lateralities**.
Note 1: A **midline** melanoma is a different laterality than right or left.
Note 2: If the laterality of one or both melanomas is **unknown**, then continue through the rules
Note 3: If one or more of the sites does not require laterality to be coded (laterality required = no in Table 1), then continue through the rules.
Example 1: Melanoma of the right side of the chest and melanoma at midline of the chest are different lateralities and are multiple primaries.

Example 2: A melanoma of the right side of the chest and a melanoma of the left side of the chest are multiple primaries.

Rule M5 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more different subtypes/variants in Column 3, **Table 2** in the Equivalent Terms and Definitions. Timing is irrelevant.

Example: Epithelioid cell melanoma 8771/3 and nodular melanoma 8721/3 are both subtypes/variants of melanoma NOS 8720/3.

Rule M6 Abstract a **single primary**[i] when synchronous, separate/non-contiguous tumors are on **the same row in Table 2** in the Equivalent Terms and Definitions. Tumors must have the same site code.

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3)

Example 1: Both lesions are located on the right side of the body and sites are chest C445 and back C445. Histology for the chest lesion is melanoma, NOS (8720/3) and the back lesion is nodular melanoma (8721/3). Abstract a single primary. Refer to illustration.

Example 2: Both lesions are located on the left leg. One lesion is spindle cell melanoma (8772/3) located on the front of the left hip C447. The other lesion is spindle cell melanoma (8772/3) located on the front of the left thigh C447. Refer to illustration.

Rule M7 Abstract **multiple primaries**[ii] when melanomas are diagnosed more than 60 days apart.

Example: An **invasive** melanoma that occurs **more than 60 days after** an **in situ** melanoma is a multiple primary.

Note 1: The purpose of this rule is to ensure that the case is counted as an incident (invasive) case when incidence data are analyzed.

Note 2: Abstract as multiple primaries even if the medical record/physician states it is recurrence or progression of disease.

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Rule M8 Abstract a **single primary**[i] when melanomas do not meet any of the above criteria.
Note: Use caution when applying this default rule. Please confirm that you have not overlooked an applicable rule.

This is the end of instructions for Multiple Melanomas

[i] Prepare one abstract. Use the histology rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.

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

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

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

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

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

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

This is a hierarchical list of source documentation.

Code the **most specific** pathology/tissue from either **resection** or **biopsy**.

Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

1. **Tissue or pathology report from primary site** (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis / synoptic report as required by CAP
   C. CAP protocol

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

Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.
Note 3: The CAP protocol is a checklist which:

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

2. Tissue/pathology from a **metastatic** site
   Note 1: Code the behavior /3.
   Note 2: The tissue from a **metastatic s**ite often shows **variations** from the primary tumor. When it is the only tissue available, it is **more accurate** than a scan.

3. **Scans: MRI, CT, PET. There is no priority** order because scans are not a reliable method for **identifying** specific **histology**(ies).

4. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following priority order:
   A. Treatment plan
   B. Documentation from Tumor Board
   C. Documentation in the medical record that refers to original pathology, cytology, or scan(s)
   D. Physician's **reference to** type of cancer (**histology**) in the medical record
      Note 1: Code the specific histology when documented.
      Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Coding Histology

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

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

   Example 1: Diagnosis for a single melanoma is melanoma, NOS 8720 with the majority or predominant part of tumor being nodular melanoma 8721. Code the subtype/variant: nodular melanoma 8721.
   Example 2: Diagnosis for a single melanoma is melanoma, NOS 8720 with the minority of tumor being amelanotic melanoma 8730. Code the subtype/variant: amelanotic melanoma 8730.
   Example 3: Diagnosis for a single tumor is melanoma, NOS 8720 with a component of malignant desmoplastic melanoma 8745. Code the subtype/variant: malignant desmoplastic melanoma 8745.

   Note 1: The terms above (A, B, C) must describe a **melanoma** in order to code a histology described by those terms.
   Example: When the diagnosis is melanoma with a nodular **melanoma** component, code nodular melanoma 8721.
   Negative Example: When the diagnosis is simply melanoma with a nodular component, code melanoma, NOS 8720. Do not assume this is a nodular melanoma.

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

2. Code the histology 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".
   Note: Do not code differentiation or features when there is no specific ICD-O code.

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

      Example: Outpatient biopsy says probably melanoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology melanoma. NOS. The case meets the criteria in #**3A.**

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

      Example 1: The pathology diagnosis is melanoma consistent with desmoplastic melanoma. The oncology consult says the patient has desmoplastic melanoma. This is clinical confirmation of the diagnosis, code desmoplastic melanoma. The case meets the criteria in **bullet 1**.

      Example 2: The pathology diagnosis is melanoma, NOS consistent with nodular melanoma. The treatment plan says the patient will receive treatment for nodular melanoma. Treatment plan confirms nodular melanoma; code nodular melanoma. The case meets the criteria in **bullet 2**.

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

List of Ambiguous Terminology

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

4. **DO NOT CODE** histology when described as:

   - Architecture
   - Foci; focus; focal
   - Pattern

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Single Melanoma or Multiple Melanomas Abstracted as a Single Primary

Rule H1 Code the histology when only **one histologic type** is identified.
Note 1: Use **Table 2** to code histology. New terms and synonyms are included in **Table 2** and coding errors may occur if the table is not used.
Note 2: When the histology is **not listed** in **Table 2,** use the **ICD-O** and all **updates**.
Note 3: When the histology includes the term regressing or regression, continue through the rules.
Note 4: When the histology includes the term lentigo maligna melanoma, continue through the rules.
Note 5: Submit a question to **Ask a SEER Registrar** when the histology code is not found in Table 2, ICD-O, or all updates.

Rule H2 Code the invasive histology when there are **invasive and in situ** components.

Rule H3 Code the **histologic type** when the diagnosis is **regressing** melanoma **and** a **histologic type**.
Example: The lesion has evidence of regressing melanoma, nodular melanoma type**.** Code 8721/3 (Nodular melanoma).

Rule H4 Code **8723/3** (Malignant melanoma, regressing) when the diagnosis is **regressing melanoma**.
Example 1*:* Malignant melanoma with regression. Code 8723 malignant melanoma, regressing.
Example 2: Malignant melanoma with features of regression. Code 8720/3 melanoma NOS.

Rule H5 Code the **histologic type** when the diagnosis is **lentigo maligna** melanoma **and** a **histologic type**.
Example**:** The diagnosis is lentigo maligna melanoma with desmoplastic melanoma, right arm. Code desmoplastic melanoma, 8745/3.

Rule H6 Code **8742/3** (Lentigo maligna melanoma) when the diagnosis is **lentigo maligna** melanoma with **no** other histologic types.

Rule H7 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS.
- Melanoma NOS 8720 and a subtype/variant of melanoma

Note*:* Use **Table 2** in the Equivalent Terms and Definitions to determine NOS and subtype/variant.

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Rule H8 Code single tumors with **two variants** as follows:

- Code **8721/3** when Nodular melanoma is mixed with:
  - Amelanotic melanoma **OR**
  - Desmoplastic melanoma **OR**
  - Epithelial cell melanoma
- Code **8730/3** when amelanotic melanoma is mixed with:
  - Spindle cell melanoma, NOS
- Code **8743/3** when Low cumulative sun damaged melanoma/superficial spreading melanoma is mixed with:
  - Desmoplastic melanoma **OR**
  - Nodular melanoma **OR**
  - Spindle cell melanoma
- Code **8744/3** when Acral melanoma/acral lentiginous melanoma, malignant is mixed with:
  - All other melanoma subtype/variants listed in Table 2
- Code **8745/3** when desmoplastic melanoma is mixed with:
  - Spindle cell melanoma, NOS

Note 1: Percentage of a subtype/variant is not used to determine histology for mixed melanomas
Note 2: If the mixed subtypes/variants are not included in this rule, continue to the next rule

Rule H9 When two or more melanoma subtype/variants are present in a single tumor and are not listed in the previous rule, submit a question to **Ask A SEER Registrar** for coding instructions.
Note 1: Two or more melanoma subtype/variants identified in a single tumor is infrequent.
Note 2: The WHO Classification of Skin Tumors 4th Ed does not include ICD-O codes for tumors with mixed melanoma subtype/variants

This is the end of instructions for Single Melanoma or Multiple Melanomas Abstracted as a Single Primary.

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

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[i] Prepare one abstract. Use the histology rules to assign the appropriate histology code. For registries collecting recurrence data: When a subsequent tumor is "single primary," record that subsequent tumor as a recurrence.
[ii] Prepare two or more abstracts. Use the histology rules to assign the appropriate histology code to each case abstracted.

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Introduction

Note 1: Tables and rules refer to ICD-O rather than ICD-O-3.2. The version is not specified to allow for updates. Use the currently approved version of ICD-O. Tables and rules refer to ICD-O rather than ICD-O-3. The version is not specified to allow for updates. Use the currently approved version of ICD-O.

Note 2: 2007 MPH Other Site Rules and 2018 Solid Tumor Other Site Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2022: Use 2007 MPH Rules
- Tumors diagnosed 01/01/2023 and later: Use the 2023 Solid Tumor Rules and Solid Tumor General Instructions
- An original tumor diagnosed ***before*** 1/1/2018 and a subsequent tumor diagnosed 1/1/2023 or later in the same primary site: Use the 2023 Solid Tumor Rules and Solid Tumor General Instructions

Note 3: For those sites/histologies which have recognized biomarkers, the biomarkers are most frequently used to target treatment. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries and/or histologic type. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Note 4: De novo (previously called frank) adenocarcinoma arises in the mucosa of the small bowel/intestines, not in a polyp.

Note 5: Polyp-specific ICD-O codes remain valid for small bowel/intestine sites.

Changes from 2007 MPH Rules

These changes are effective with cases diagnosed 1/1/2023 and later. Changes are based on 4th and 5th Edition WHO Classification of Tumors Books for the following sites: Digestive System Tumors, Female Genital Tumors, Endocrine Organs, Tumors of the Eye, Soft Tissue and Bone, and Urinary and Male Genital Organs.

1. The previous 2007 MPH Rules instructed you to "Code the histology from the most representative specimen." For all sites included in 2023 Other Sites Solid Tumor Rules, the instruction is now "Code the most specific histology from biopsy or

resection. When there is a discrepancy between the biopsy and resection (two distinctly different histologies), code the histology from the most representative specimen (the greater amount of tumor)."

2. Histology tables for the majority of site groups covered by Other Sites Solid Tumor Rules have been added as histology coding reference tools. See the Site or Site Group Histology-Specific Tables section for more information.

3. 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.
   Note 1: Not all sites are included in the tables
   Note 2: Each histology table may include coding tips specific to that site group.
   Note 3: To assign the correct ICD-O code, it is necessary to refer to the site-specific histology table to determine if there are additional coding instructions or criteria that must be met to assign a code.
   Note 4: Given the number of sites included in Other Sites Rules, additional histology coding (H) rules were limited to the more common sites.

4. Rectum and Rectosigmoid were included in the Colon Rules beginning 1/1/2018.

5. Bilateral epithelial ovarian tumors must be the same histology or be an NOS and subtype/variant in order to be coded as a single primary beginning 1/1/2023.

6. Paraganglioma histologies 8680/3, 8690/3, 8692/3, and 9693/3 for primary sites C479, C754 and C755 ONLY are in the Head and Neck module (Table 9) for cases diagnosed 1/1/2019 forward. All other histologies (except for hematopoietic and lymphoid), and paraganglioma histologies before 2019 should use Other Sites.

New for 2024

1. Guidelines for assigning primary sites for liver and intrahepatic bile duct neoplasms based on histology and other criteria are included in the newly added Table 9a. The criteria for coding liver (C220) versus intrahepatic bile duct (C221) is based on Cancer PathCHART Specialty Matter Expert review. The experts have determined adenocarcinoma and subtypes of adenocarcinoma cannot be primary to liver and therefore are biologically impossible. The coding instructions in Table 9a may be applied to cases diagnosed 2023 forward.

2. Several tables in the Solid Tumor Other Sites module include more than one site or site group. The tables are based on WHO Classifications of Tumors books unless otherwise noted. The Cancer PathCHART review determined that some histologies are valid for specific sites only and not for all sites within a site group. The valid C-code will be denoted in bold next to the histology or histologies in applicable tables. Coding these histologies to a site other than the one(s) noted in the tables has been determined to be biologically impossible and will not pass edits.

Equivalent or Equal Terms

These terms can be used interchangeably:

- Acinar adenocarcinoma, adenocarcinoma (for prostate only)
- Adenocarcinoma, glandular carcinoma
- And; with; (duct **and** lobular is equivalent to duct **with** lobular)
  Note: "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Basal cell carcinoma; basal cell adenocarcinoma (Prostate primaries only, both are coded 8147)
- Carcinoid; NET; neuroendocrine tumor
- Carcinoma; adenocarcinoma
  - A histology type must be stated for these terms to be equal
  - Example: Serous carcinoma and serous adenocarcinoma are both coded 8441
- Contiguous; continuous
- In situ; noninvasive; intraepithelial
- Multicentric; multifocal
- Mucinous; mucoid; mucous; colloid
- Neuroendocrine carcinoma; NEC
- Polyp; adenoma; polyp NOS; adenomatous polyp
- Serosa; visceral peritoneum
- Simultaneous; existing at the same time; concurrent; prior to first course treatment
- Site; topography
- Tumor; mass; tumor mass; lesion; neoplasm

  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician's statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to **determine** multiple primaries
  - **Do not** use these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

Terms that are NOT Equivalent or Equal

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

- **Bilateral** is not equivalent to either **single primary** or **multiple primaries.** See Multiple Primary rules for instructions.
- **Carcinoma, NOS 8010** is not equivalent to **adenocarcinoma, NOS 8140**
- **Component** is not equivalent to **subtype/type/variant**
  - **Note:** Component is only coded when the pathologist specifies the component as a second carcinoma/sarcoma
- **Phenotype** is not equivalent to **subtype/type/variant**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Site or Site Group Histology-Specific Tables

Twenty-one site-specific histology tables are included in the Solid Tumor Other Sites module. Each table applies to a site or site group and lists histologies that commonly occur in those sites. These tables are based on the most recent WHO Classification of Tumors Books and/or College of American Pathologist (C.A.P.) protocols and do not list all possible histologies that may arise in that site.

In place of adding numerous site-based histology rules to the Other Sites module, 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.

Coding instructions and/or helpful information are located above the tables. Additional notes are found next to specific histologies listed in the table columns.

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

Table Index

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Table Number	Table Title
Table 9a	Guidelines for Assigning Primary Site for Liver and Intrahepatic Bile Duct **C220**, **C221**
Table 10	Gallbladder and Extrahepatic Bile Ducts Histologies **C239, C240, C248**, **C249**
Table 11	Pancreas Histologies **C250-C254, C257, C258, C259**
Table 12	Thyroid Histologies **C739**
Table 13	Ovary Histologies **C569**
Table 14	Peritoneum Histologies **C482**
Table 15	Fallopian Tube Histologies **C570**
Table 16	Uterine Corpus Histologies **C540-C543, C548, C549, C559**
Table 17	Uterine Cervix Histologies **C530-C531, C538, C539**
Table 18	Vagina Histologies **C529**
Table 19	Vulva Histologies **C510-C512, C518, C519**
Table 20	Soft Tissue Histologies **C490-C496, C498, C499**
Table 21	Bone Histologies **C400-C403, C408, C409, C412-C414, C418, C419**
Table 22	Thymus Histologies **C379**
Table 23	Penis and Scrotum Histologies **C600-C602, C608, C609, C632**

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Table 1: Paired Organs and Sites with Laterality

Laterality must be coded for all of the following sites. SEER does allow coding laterality for sites not listed in Table 1.

Site Code	Site or Subsite
C384	Pleura
C400	Long bones of upper limb, scapula, and associated joints
C401	Short bones of upper limb and associated joints
C402	Long bones of lower limb and associated joints
C403	Short bones of lower limb and associated joints
C413	Rib, clavicle (excluding sternum)
C414	Pelvic bones (excluding sacrum, coccyx, symphysis pubis)
C441	Skin of the eyelid
C442	Skin of the external ear
C443	Skin of other and unspecific parts of the face (if midline, assign code 5)
C444	Skin of scalp and neck
C445	Skin of the trunk (if midline, assign code 5)
C446	Skin of upper limb and shoulder
C447	Skin of the lower limb and hip
C471	Peripheral nerves and autonomic nervous system of upper limb and shoulder
C472	Peripheral nerves and autonomic nervous system of the lower limb and hip
C491	Connective, subcutaneous, and other soft tissues of upper limb and shoulder

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Site Code	Site or Subsite
C492	Connective, subcutaneous, and other soft tissues of the lower limb and hip
C569	Ovary
C570	Fallopian tube
C620-C629	Testis
C630	Epididymis
C631	Spermatic cord
C690-C699	Eye and adnexa
C740-C749	Adrenal gland

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Table 2: Mixed and Combination Codes

Instructions:

1. Compare the **terms** in the **diagnosis** (pathology, cytology, radiographic, clinical) to the terms in **Column 1.**
2. When the terms **match**, use the **combination code** listed in **Column 2.**
3. The **last row** in the table is a **"last resort"** code: adenocarcinoma mixed subtypes 8255.
4. Do not use this table unless instructed to by the Histology Rules.

IMPORTANT NOTE: Histology Tables 3-23 may include additional coding instructions for "mixed" histologies.

Note 1: **Do not** use Table 2 in the following situations:

- For tumors with both **invasive** and **in situ** behavior. The **Histology Rules** instruct to code the invasive histology.
- When one of the histologies is described as **differentiation or features.** A histology with differentiation or features is a single histology.
- When the terms are a **NOS** and a **subtype/variant** of that NOS. See the **Histology Rules** for instructions on coding a NOS and a subtype/variant in a single tumor or multiple tumors abstracted as a single primary.

Note 2: Some combinations can be either in situ or invasive; others are limited to a /2 or /3 behavior code.

- When a code is **limited to in situ, /2** will be **added** to the code (both components are in situ)
- When a code is **limited to invasive, /3** will be **added** to the code (both components are invasive)

Note 3: This table is not a complete listing of histology combinations.

Column 1 lists the **required terms** for **the combination code.**
Column 2 lists the **combination term** and **code** for histologies **in Column 1.**

Table begins on next page.

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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**
Large cell neuroendocrine carcinoma **AND** Adenocarcinoma NOS **OR** Squamous cell carcinoma NOS **OR** Spindle cell carcinoma **OR** Giant cell carcinoma	Combined large cell neuroendocrine carcinoma **8013**

Other Sites Equivalent Terms and Definitions
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Required Histology Terms	Histology Combination Term and Code
Squamous carcinoma **AND** Basal cell carcinoma	Basosquamous carcinoma **8094**
Islet cell **AND** Exocrine carcinoma	Mixed islet cell and exocrine adenocarcinoma **8154**
Acinar **AND** Endocrine/neuroendocrine	Mixed acinar-endocrine/neuroendocrine carcinoma **8154**
Acinar **AND** **Both of the following**: Endocrine Ductal	Mixed acinar-endocrine-ductal carcinoma **8154**
Ductal **AND** Endocrine/neuroendocrine	Mixed ductal-endocrine carcinoma **8154** Mixed ductal-neuroendocrine carcinoma **8154**

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Required Histology Terms	Histology Combination Term and Code
Endocrine **AND** Exocrine	Mixed endocrine and exocrine adenocarcinoma **8154**
Hepatocellular carcinoma **AND** Cholangiocarcinoma	Combined hepatocellular carcinoma and cholangiocarcinoma **8180 (C221)**
Adenocarcinoma **AND** Carcinoid/neuroendocrine carcinoma(NEC)/neuroendocrine tumor (NET)	Mixed adenoneuroendocrine carcinoma/combined carcinoid and adenocarcinoma **8244**
Adenocarcinoma **AND** **At least two** of the following: Papillary Clear cell Mucinous/colloid Signet ring Acinar	Adenocarcinoma with mixed subtypes/Adenocarcinoma combined with other types of carcinoma **8255** ***Note:*** Code 8255 does not apply to GYN primaries. Continue through the table to determine correct mixed histology code for GYN neoplasms**.**

Other Sites Equivalent Terms and Definitions
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Required Histology Terms	Histology Combination Term and Code
Gyn malignancies with **two or more** of the following: Clear cell Endometrioid Mucinous Papillary Serous Squamous	Mixed cell adenocarcinoma **8323** ***Note:*** First refer to ICD-O-3.2 and ICD-O updates to confirm if the mixed histology has a specific code. ***Example*:** Serous papillary adenocarcinoma is coded 8441 per ICD-O-3.2.
Papillary thyroid carcinoma (includes subtype/variants) **AND** Follicular (includes subtype/variants)	Papillary carcinoma, follicular variant **8340** (synonyms: Infiltrative follicular variant of papillary carcinoma; Widely invasive follicular variant of papillary carcinoma) ***Note:*** First refer to ICD-O-3.2 and ICD-O updates to confirm if the mixed histology has a specific code.
Medullary **AND** Follicular (includes subtype/variants)	Mixed medullary-follicular carcinoma **8346**
Medullary **AND** Papillary (includes subtype/variants)	Mixed medullary-papillary carcinoma **8347**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
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Required Histology Terms	Histology Combination Term and Code
Medullary **AND** Poorly differentiated carcinoma **OR** Anaplastic carcinoma **OR** Oncocytic carcinoma	Mixed medullary and poorly differentiated carcinoma / mixed medullary and anaplastic carcinoma / Mixed medullary and oncocytic carcinoma **8346**
Squamous carcinoma **AND** Adenocarcinoma	Adenosquamous carcinoma **8560**
Any combination of the following sarcomas: Myxoid Round cell Pleomorphic	Mixed liposarcoma **8855**
Embryonal rhabdomyosarcoma **AND** Alveolar rhabdomyosarcoma	Mixed type rhabdomyosarcoma **8902**
Teratoma **AND** Embryonal carcinoma	Teratocarcinoma **9081**

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Required Histology Terms	Histology Combination Term and Code
Any combination of the following: Embryonal carcinoma Seminoma Teratoma Yolk sac tumor	Mixed germ cell tumor **9085**
Choriocarcinoma **AND** **Any of the following:** Embryonal Seminoma Teratoma	Choriocarcinoma combined with other germ cell elements **9101**

Table 3: Prostate Histologies

Table 3 lists the more common histologies for prostate.
C619 Prostate gland; prostate, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

- Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, acinar adenocarcinoma NOS 8140/3 (column 1) is a generic term which encompasses a number of histologies, including ductal adenocarcinoma 8500/3 (column 3). Ductal adenocarcinoma is also a NOS because it has subtypes/variants. The subtypes/variants are indented under the NOS (ductal adenocarcinoma) in column 3. There is also a note in column 1 which calls attention to the fact that ductal adenocarcinoma has subtypes/variants.
    - When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, ductal adenocarcinoma **8500/3** and papillary adenocarcinoma **8260/3** are a NOS and a subtype/variant, **NOT** two different subtypes.

Continued on next page

Other Sites Equivalent Terms and Definitions
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Coding notes for acinar adenocarcinoma subtype/variants:

- **Ductal adenocarcinoma 8500/3:** In prostate biopsies, the term "adenocarcinoma of prostate with ductal features" should be used in the pathology report and is coded 8140/3. In order to code ductal adenocarcinoma 8500/3, the ductal component must comprise >50% of the tumor with the percentage reported and from a radical prostatectomy specimen.
- **Intraductal carcinoma of prostate 8500/2:** Intraductal prostate carcinoma is most often associated with invasive acinar adenocarcinoma of ductal carcinoma.
- **Mucinous adenocarcinoma 8480/3:** In order to code 8480/3, the mucinous adenocarcinoma component must comprise >25% of the tumor, so the diagnosis must be made only in excision specimens.
- **Sarcomatoid carcinoma 8572/3:** Exceedingly rare and most commonly occurs during the development of high-grade adenocarcinoma, especially after irradiation.
- **Signet ring cell-like adenocarcinoma 8490/3:** In order to code 8490/3, the signet-ring-like cells must comprise >25% of tumor, so the diagnosis must be made only in excision specimens.

Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Acinar adenocarcinoma **8140** ***Note:*** Ductal/intraductal adenocarcinoma 8500 is also a NOS with the following subtypes/variants: Cribriform adenocarcinoma **8201/3** Papillary adenocarcinoma **8260/3** Solid adenocarcinoma **8230/3**	Acinar carcinoma Adenocarcinoma in situ **8140/2** Adenocarcinoma, NOS **8140/3** Adenocarcinoma with ductal features **8140/3** Atrophic adenocarcinoma **8140/3** Foamy gland adenocarcinoma **8140/3** Microcystic adenocarcinoma **8140/3** Pseudohyperplastic adenocarcinoma **8140/3** Prostatic intraepithelial-like carcinoma **8140/3**	Acinar adenocarcinoma, sarcomatoid variant **8572** Ductal/intraductal adenocarcinoma **8500** Cribriform adenocarcinoma **8201** Papillary adenocarcinoma **8260** Solid adenocarcinoma **8230** Mucinous (colloid) adenocarcinoma **8480** Signet ring-like cell adenocarcinoma **8490**

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Adenocarcinoma with neuroendocrine differentiation **8574/3** ***Note 1:*** This histology is considered treatment-related neuroendocrine prostatic carcinoma demonstrating complete neuroendocrine differentiation or partial neuroendocrine differentiation with adenocarcinoma after androgen-deprivation therapy. ***Note 2*:** Code 8574/3 only when there is no history of previous prostate adenocarcinoma or history of androgen-deprivation therapy.		
Adenosquamous carcinoma **8560**	Prostatic carcinoma with adenosquamous differentiation	
Basal cell adenocarcinoma **8147**	Adenoid cystic basal cell carcinoma Adenoid cystic carcinoma Adenoid cystic carcinoma (solid pattern) Basal cell carcinoma of prostate	
Mixed acinar-ductal adenocarcinoma **8552** ***Note*:** Assign code 8552 when the ductal component is not stated or less than 50% of the tumor.		

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Neuroendocrine tumor **8240/3** ***Note 1:*** 50% of SmCC of prostate cases present as a de novo malignancy ***Note 2:*** SmCC of the prostate often occurs following androgen deprivation treatment for acinar adenocarcinoma	Well differentiated neuroendocrine tumor WD neuroendocrine tumor	Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Sarcoma, NOS **8800/3**	Mesenchymal tumor, malignant	Stromal sarcoma **8935/3** Leiomyosarcoma **8890/3** Rhabdomyosarcoma **8900/3** Angiosarcoma **9120/3** Synovial sarcoma **9040/3** Osteosarcoma **9180/3** Undifferentiated pleomorphic sarcoma **8802/3** Solitary fibrous tumor, malignant **8815/3**
Squamous cell carcinoma **8070** ***Note*:** In >50% of reported cases, there is an association with previous hormone or radiation therapy for prostatic adenocarcinoma. If a patient has a known history of acinar adenocarcinoma of prostate treated with hormone and/or radiation and subsequent findings of SCC, this is recurrence and not a new primary.	SCC, NOS	

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Urothelial carcinoma **8120** ***Note 1:*** Primary urothelial carcinoma of the prostate can rarely occur in the absence of a bladder tumor. ***Note 2***: Urothelial carcinoma of the prostate are almost always found in the prostatic urethra.		

Other Sites Equivalent Terms and Definitions
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Table 4: Testis Histologies

Table 4 lists the more common histologies for testis as stated in the College of American Pathologists (C.A.P.) testis protocol
C620 Undescended testis
C621 Descended testis
C629 Testis, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

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Other Sites Equivalent Terms and Definitions
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Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Germ cell tumor NOS **9064** ***Note 1:*** The following teratomas are not **reportable:** • Teratoma, prepubertal type 9084/0 • Teratoma, mature, prepubertal type 9084/0 ***Note 2:*** The following yolk sac tumor patterns are all coded 9071: • Endodermal sinus/perivascular pattern • Glandular/alveolar pattern • Hepatoid pattern • Macrocystic pattern • Microcystic/reticular pattern • Myxoid pattern • Papillary pattern • Parietal pattern • Polyvesicular vitelline pattern • Sarcomatoid/spindle cell pattern • Solid pattern	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 (see Note 2)** Teratoma with malignant transformation/Teratoma with somatic-type malignancy **9084**
Leydig cell tumor, malignant **8650/3**		
Seminoma, NOS **9061**	Seminoma with syncytiotrophoblastic cells	
Sertoli cell carcinoma **8640/3**	Sertoli cell tumor, malignant	

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Table 5: Esophagus Histologies

Table 5 list the more common histologies for the following esophagus subsites:
C150 Cervical esophagus
C151 Thoracic esophagus
C152 Abdominal esophagus
C153 Upper third of esophagus (proximal third of esophagus)
C154 Middle third of esophagus
C155 Lower third of esophagus (Distal third of esophagus)
C158 Overlapping lesion of esophagus
C159 Esophagus, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Adenocarcinoma, NOS **8140**	Adenocarcinoma in situ **8140/2**	
Adenoid cystic carcinoma **8200**		
Adenosquamous carcinoma **8560**		
Gastrointestinal stromal tumor **8936**	GANT Gastrointestinal pacemaker cell tumor Gastrointestinal stromal tumor GIST, NOS GIST, malignant Gastrointestinal stromal sarcoma Succinate dehydrogenase-deficient gastrointestinal stromal tumor	
Mucoepidermoid carcinoma **8430**		
Squamous cell carcinoma **8070**	Squamous carcinoma Squamous cell carcinoma in situ **8070/2** Squamous cell carcinoma, usual type	Basaloid squamous cell carcinoma **8083** Squamous cell carcinoma, spindle cell/squamous cell carcinoma, sarcomatoid **8074** Verrucous squamous cell carcinoma **8051**
Undifferentiated carcinoma **8020/3**		
Neuroendocrine tumor **8240/3**	NET	Neuroendocrine carcinoma **8246/3** Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Mixed neuroendocrine-non-endocrine neoplasm (MiNEN) **8154/3** ***Note*:** Esophageal MiNENs usually consist of poorly differentiated NEC and either squamous cell carcinoma or adenocarcinoma	MiNEN	

Other Sites Equivalent Terms and Definitions
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Table 6: Stomach Histologies

Table 6 list the more common histologies for the following stomach subsites:
C160 Cardia, NOS; gastric cardia; cardioesophageal junction; esophagogastric junction; gastroesophageal junction
C161 Fundus of stomach; gastric fundus
C162 Body of stomach; corpus of stomach; gastric corpus
C163 Gastric antrum; antrum of stomach; pyloric antrum
C164 Pylorus; pyloric canal; prepylorus
C165 Lesser curvature of stomach, NOS
C166 Greater curvature of stomach, NOS
C168 Overlapping lesion of stomach
C169 Stomach, NOS; gastric, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

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

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Coding notes for Stomach:
- When both Lauren and WHO histologies are stated, code the WHO diagnosis.
  - Exception: If WHO diagnosis is adenocarcinoma, NOS and Lauren indicates a more specific type, code the specific histology.

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Adenocarcinoma, NOS **8140** ***Note 1*:** For stomach sites, code mucinous carcinoma (8480) or signet-ring cell carcinoma (8490) regardless of percentage. ***Note 2***: Paneth cell carcinoma is a rare subtype of adenocarcinoma. A specific ICD-O code has not been proposed by WHO. Code as 8140.	Adenocarcinoma in situ **8140/2** Adenocarcinoma of fundic gland type Paneth cell carcinoma	Adenocarcinoma, diffuse type **8145/3** Adenocarcinoma, intestinal type **8144/3;** Intestinal type adenoma, high grade **8144/2** Hepatoid adenocarcinoma **8576** Medullary carcinoma with lymphoid stroma **8512** Micropapillary carcinoma **8265** Mucinous adenocarcinoma **8480** Mucoepidermoid carcinoma **8430** Papillary adenocarcinoma **8260** Parietal cell carcinoma **8214** Signet ring cell carcinoma/Poorly cohesive carcinoma **8490** Tubular adenocarcinoma **8211**
Adenomatous polyp, high grade **8210/2**	Adenomatous polyp, high grade dysplasia	
Adenosquamous carcinoma **8560**		
Gastroblastoma **8976/3**		
Gastrointestinal stromal tumor **8936**	GANT Gastrointestinal pacemaker cell tumor Gastrointestinal stromal tumor GIST, NOS GIST, malignant Gastrointestinal stromal sarcoma Succinate dehydrogenase-deficient gastrointestinal stromal tumor	
Glandular intraepithelial neoplasia, high grade **8148/2**	Glandular intraepithelial neoplasia, grade III	

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Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Mixed adenoneuroendocrine carcinoma **8244/3**	Combined carcinoid and adenocarcinoma Composite carcinoid MANEC Mixed carcinoid and adenocarcinoma	
Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN) **8154/3**		
Neuroendocrine carcinoma (NEC) **8246/3**		Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor, NOS **8240/3**	Carcinoid Neuroendocrine tumor, grade 1 Well differentiated endocrine tumor/carcinoma	Enterochromaffin-like cell tumor **8242/3** Neuroendocrine tumor, EC-cell, serotonin-producing **8241/3** Neuroendocrine tumor, gastrin-producing (gastrinoma) **8153/3** Neuroendocrine tumor grade 2/neuroendocrine tumor grade 3 **8249/3**
Serrated dysplasia, high grade **8213/2**		
Squamous cell carcinoma **8070**		
Undifferentiated carcinoma **8020/3**		Carcinoma with osteoclast-like giant cells **8035/3** Large cell carcinoma with rhabdoid phenotype **8014/3** Pleomorphic carcinoma **8022/3** Sarcomatoid carcinoma **8033/3**

Other Sites Equivalent Terms and Definitions
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Table 7: Small Intestine and Ampulla of Vater Histologies

Table 7 list the more common histologies for the following small intestine subsites:
C170 Duodenum
C171 Jejunum
C172 Ileum (excludes ileocecal valve C180)
C173 Meckel diverticulum
C178 Overlapping lesion of small intestine
C179 Small intestine, NOS; small bowel, NOS
C241 Ampulla of Vater; periampullary

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Adenocarcinoma **8140**	Ampullary carcinoma	Adenocarcinoma, intestinal type **8144/3;** Intestinal type adenoma, high grade **8144/2** Medullary adenocarcinoma **8510** Mucinous adenocarcinoma **8480** Non-invasive pancreatobiliary papillary neoplasm with high grade dysplasia **8163/2;** Pancreatobiliary-type carcinoma **8163/3** Poorly cohesive carcinoma/signet-ring cell carcinoma **8490** Tubular adenocarcinoma **8211**
Adenomatous polyp, high grade **8210/2**	Adenomatous polyp, high grade dysplasia	
Gastrointestinal stromal tumor **8936**	GANT Gastrointestinal pacemaker cell tumor Gastrointestinal stromal tumor GIST, NOS GIST, malignant Gastrointestinal stromal sarcoma Succinate dehydrogenase-deficient gastrointestinal stromal tumor	
Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN) **8154/3**		
Neuroendocrine carcinoma **8246/3**		Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonym	Subtypes/Variants
Neuroendocrine tumor **8240/3**	Neuroendocrine tumor, grade 1	Neuroendocrine tumor, grade 2/neuroendocrine tumor, grade 3 **8249/3**
Serrated dysplasia, high grade **8213/2**		

Table 8: Anus Histologies

Table 8 list the more common histologies for the following anal subsites:
C210 Anus, NOS
C211 Anal canal; anal sphincter
C212 Cloacogenic zone
C218 Overlapping lesion of rectum, anus, and anal canal

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Coding Notes for Anus: p16 test results can be used to code squamous cell carcinoma, HPV positive (8085) and squamous cell carcinoma, HPV negative (8086).

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma **8140**		
Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN) **8154/3**		
Neuroendocrine carcinoma **8246/3**		Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor **8240/3**	Neuroendocrine tumor, grade 1	Neuroendocrine tumor, grade 2/neuroendocrine tumor, grade 3 **8249/3**
Squamous cell carcinoma **8070**	Squamous cell carcinoma, usual type	Squamous cell carcinoma, HPV negative **8086** Squamous cell carcinoma, HPV positive **8085** Verrucous squamous cell carcinoma **8051**
Squamous intraepithelial neoplasia, high grade **8077/2**	AIN, grade II AIN, grade III Anal intraepithelial neoplasia, grade II Anal intraepithelial neoplasia, grade III HSIL Squamous intraepithelial neoplasia, grade II Squamous intraepithelial neoplasia, grade III	

Table 9: Liver and Intrahepatic Bile Duct Histologies

Table 9 list the more common histologies for the following liver and intrahepatic bile duct subsites:
C220 Liver; hepatic, NOS
C221 Intrahepatic bile duct; biliary canaliculus; cholangiole

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Cases diagnosed before 1/1/2023:

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.

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonyms	Subtypes/Variants
Carcinoma, undifferentiated **8020/3**		
Cholangiocarcinoma **8160/3 (C221)**	Bile duct adenocarcinoma/carcinoma Intrahepatic cholangiocarcinoma (iCCA) Large duct intrahepatic cholangiocarcinoma Small duct intrahepatic cholangiocarcinoma	
Combined hepatocellular carcinoma and cholangiocarcinoma **8180/3 (C221)**	Hepatocholangiocarcinoma Mixed hepatobiliary carcinoma Mixed hepatocellular-cholangiocarcinoma	
Hepatoblastoma **8970/3**		
Hepatocellular carcinoma **8170/3 (C220)**	Hepatocarcinoma Hepatoma, malignant Hepatoma, NOS	Hepatocellular carcinoma, fibrolamellar **8171** Hepatocellular carcinoma, scirrhous / sclerosing hepatic carcinoma **8172** Hepatocellular carcinoma (HCC), clear cell **8174** (synonyms below) HCC, chromophobe HCC, lymphocytic-rich HCC, macrotrabecular massive HCC, neutrophile-rich HCC, steatohepatitic
Intraductal papillary neoplasm **8503**	Intraductal papillary neoplasm with associated invasive carcinoma **8503/3** Intraductal papillary neoplasm with high grade intraepithelial neoplasia **8503/2**	
Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN) **8154/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonyms	Subtypes/Variants
Mucinous cystic neoplasm **8470**	Mucinous cystic neoplasm with associated invasive carcinoma **8470/3** Mucinous cystic neoplasm with high grade intraepithelial neoplasia **8470/2**	
Neuroendocrine carcinoma **8246/3**		Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor **8240/3**	Neuroendocrine tumor, grade 1	Neuroendocrine tumor, grade 2/ neuroendocrine tumor, grade 3 **8249/3**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Table 9a: Guidelines for Assigning Primary Site for Liver and Intrahepatic Bile Duct

C220 Liver; hepatic, NOS
C221 Intrahepatic bile duct; biliary canaliculus; cholangiole

Guidelines for assigning primary sites for liver and intrahepatic bile duct neoplasms based on histology and other criteria are included in the newly added Table 9a. The criteria for coding liver (C220) versus intrahepatic bile duct (C221) is based on Cancer PathCHART Specialty Matter Expert review. The experts have determined adenocarcinoma and subtypes of adenocarcinoma cannot be primary to liver and therefore are biologically impossible. This table may be applied to cases diagnosed 2023 forward.

Column 1 contains the site of the biopsy specimen and/or cytology specimen
Column 2 contains the histology diagnosis as stated by the pathologist
Column 3 contains the criteria required to assign primary site based on Cancer PathCHART Specialty Matter Expert review
Column 4 contains the primary site and histology to be assigned

Site of biopsy or cytology	Pathology or cytology diagnosis	Criteria	Primary Site/ Histology
Liver C220	Adenocarcinoma Adenocarcinoma subtypes/variants	Supporting documentation such as scans, lab tests, or definitive clinical diagnosis of intrahepatic bile duct primary and/or definitive diagnosis of cholangiocarcinoma	**C221** **8160/3**
Liver C220	Adenocarcinoma Adenocarcinoma, subtypes/variants	No documentation supporting the primary site of intrahepatic bile duct is available in the medical record. This includes scans, lab tests or definitive clinical diagnosis. Liver is a common metastatic site for other neoplasms such as breast, lung, and colon. Code unknown primary site C809 when a primary site is not indicated in the pathology report or medical record.	**C809** **8140/3**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Site of biopsy or cytology	Pathology or cytology diagnosis	Criteria	Primary Site/ Histology
Liver C220 or Intrahepatic bile ducts C221	Hepatocellular carcinoma	Cancer PathCHART review has determined hepatocellular carcinoma is valid for liver C220 only. Code C220 regardless of biopsy/cytology site.	**C220** **8170/3**
Liver C220	Combined hepatocellular carcinoma and cholangiocarcinoma	Cancer PathCHART review has determined combined hepatocellular carcinoma and cholangiocarcinoma is valid for intrahepatic bile ducts C221 only. Code C221 regardless of biopsy/cytology site	**C221** **8180/3**

Table 10: Gallbladder and Extrahepatic Bile Duct Histologies

Table 10 list the more common histologies for the following gallbladder and extrahepatic bile duct subsites:
C239 Gallbladder
C240 Extrahepatic bile duct; bile duct, NOS; biliary duct, NOS; choledochal duct; common bile duct; common duct; cystic bile duct; cystic duct; hepatic bile duct; hepatic duct; sphincter of Oddi
C248 Overlapping lesion of biliary tract
C249 Biliary tract, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

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

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma **8140**	Biliary-type adenocarcinoma **8140**	Adenocarcinoma, intestinal type **8144** Clear cell adenocarcinoma **8310** Mucinous adenocarcinoma **8480** Pancreatobiliary-type carcinoma **8163** Poorly cohesive carcinoma/signet ring cell carcinoma **8490**
Adenosquamous carcinoma **8560**		
Bile duct carcinoma **8160 (C240)**	Cholangiocarcinoma	Bile duct cystadenocarcinoma **8161** Perihilar cholangiocarcinoma **8162**
Biliary intraepithelial neoplasia, high grade **8148/2**		
Carcinoma, NOS **8010**		Undifferentiated carcinoma **8020**
Intracystic papillary neoplasm **8503**	Intracystic papillary neoplasm with high grade intraepithelial neoplasia **8503/2** Intracystic papillary tumor with high grade dysplasia **8503/2** Intraductal papillary neoplasm with high grade dysplasia **8503/2** Intraductal papillary neoplasm with high grade intraepithelial neoplasia **8503/2** Intracystic papillary neoplasm with associated invasive carcinoma **8503/3** Intraductal papillary neoplasm with associated invasive carcinoma **8503/3**	
Mixed neuroendocrine-non-neuroendocrine neoplasm (MiNEN) **8154/3**		
Mucinous cystic neoplasm with invasive carcinoma **8470/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific or NOS Terms and Code	Synonyms	Subtypes/Variants
Neuroendocrine carcinoma **8246/3**		Large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor **8240/3**	Neuroendocrine tumor, grade 1	Neuroendocrine tumor, grade 2/neuroendocrine tumor, grade 3 **8249/3**
Squamous cell carcinoma **8070**		

Table 11: Pancreas Histologies

Table 11 list the more common histologies for the following pancreas subsites:
C250 Head of pancreas
C251 Body of pancreas
C252 Tail of pancreas
C253 Pancreatic duct; duct of Santorini; duct of Wirsung
C254 Islet of Langerhans; islands of Langerhans; endocrine pancreas
C257 Other specified parts of pancreas; neck of pancreas
C258 Overlapping lesion of pancreas
C259 Pancreas, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma, NOS **8140**		Acinar cell carcinoma **8550** Colloid carcinoma/mucinous carcinoma **8480** Ductal adenocarcinoma/pancreatic ductal adenocarcinoma **8500** Hepatoid carcinoma **8576** Invasive micropapillary carcinoma **8265** Medullary carcinoma **8510** Mixed acinar-ductal carcinoma **8552** Mixed acinar neuroendocrine carcinoma/ mixed acinar-ductal neuroendocrine carcinoma **8154/3** Signet-ring cell (poorly cohesive) carcinoma **8490**
Adenosquamous carcinoma **8560**		
Glandular intraepithelial neoplasia, high grade **8148/2**	Intestinal pancreatic intraepithelial neoplasia Oncocytic pancreatic intraepithelial neoplasia Pancreatic intraepithelial neoplasia (PanIN)	
Intraductal oncocytic papillary neoplasm **8455**	Intraductal oncocytic papillary neoplasm with associated invasive carcinoma **8455/3** Intraductal oncocytic papillary neoplasm, NOS **8455/2**	

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Intraductal papillary mucinous neoplasm **8453**	Intraductal papillary mucinous neoplasm with high grade-dysplasia **8453/2** High-grade IPMN **8453/2** Intraductal papillary mucinous carcinoma, non-invasive **8453/2** Intraductal papillary mucinous carcinoma, invasive **8453/3** Intraductal papillary mucinous neoplasm with associated invasive carcinoma **8453/3**	
Intraductal tubulopapillary neoplasm **8503**	Intraductal tubulopapillary neoplasm **8503/2** Intraductal tubulopapillary neoplasm with associated invasive carcinoma **8503/3**	
Mixed neuroendocrine non-neuroendocrine neoplasm **8154**	MiNEN Mixed acinar-endocrine carcinoma Mixed acinar-endocrine-ductal carcinoma Mixed acinar-neuroendocrine carcinoma	
Mucinous cystic neoplasm **8470**	Mucinous cystic neoplasm with high-grade dysplasia **8470/2** Mucinous cystadenocarcinoma, non-invasive **8470/2** Mucinous cystic neoplasm with high grade intraepithelial neoplasia **8470/2** Mucinous cystic tumor with high grade dysplasia **8470/2** Mucinous cystic neoplasm with an associated invasive carcinoma **8470/3**	

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Neuroendocrine carcinoma, NOS **8246**	PanNEC	Large cell neuroendocrine carcinoma **8013** Small cell neuroendocrine carcinoma **8041**
Neuroendocrine tumor, NOS **8240** ***Note:*** Pancreatic neuroendocrine tumor, non-functioning has the following synonyms (they are not subtype/variants): • Clear cell neuroendocrine tumor, non-functioning pancreatic • Cystic neuroendocrine tumor, non-functioning pancreatic • Oncocytic neuroendocrine tumor, non-functioning pancreatic • Pleomorphic neuroendocrine tumor, non-functioning pancreatic	Neuroendocrine tumor, grade 1 PanNET	ACTH-producing tumor **8158** Enterochromaffin-cell carcinoid / Serotonin-producing tumor **8241** Gastrinoma **8153** Glucagonoma **8152** Insulinoma **8151** Neuroendocrine tumor grade 2 / neuroendocrine tumor grade 3 **8249** Pancreatic neuroendocrine tumor, non-functioning **8150** (see note for synonyms) Somatostatinoma **8156** VIPoma **8155**
Pancreatoblastoma **8971/3**		
Solid pseudopapillary neoplasm of pancreas **8452**	Solid pseudopapillary carcinoma Solid pseudopapillary neoplasm with high-grade carcinoma **8452/3**	
Squamous cell carcinoma **8070**		
Undifferentiated carcinoma **8020/3**		Undifferentiated carcinoma with osteoclast-like giant cells **8035/3** Undifferentiated carcinoma with rhabdoid cells **8014/3**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Table 12: Thyroid Histologies

Table 12 list the more common histologies for thyroid:
C739 Thyroid gland; thyroid, NOS; thyroglossal duct

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Thyroid Coding Note: Papillary carcinoma, follicular variant, encapsulated/well demarcated, non-invasive is not reportable.

Table begins on next page.

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Carcinoma, anaplastic **8021/3**		Carcinoma, undifferentiated **8020/3**
Follicular thyroid carcinoma, NOS **8330**	Follicular adenocarcinoma Follicular carcinoma Follicular carcinoma, widely invasive **8330/3** Infiltrative follicular carcinoma **8330/3**	Follicular carcinoma, encapsulated angioinvasive **8339/3** Follicular thyroid carcinoma, minimally invasive **8335/3** Well differentiated follicular adenocarcinoma **8331** Moderately differentiated follicular adenocarcinoma/ trabecular follicular carcinoma **8332**
Medullary thyroid carcinoma **8345**	C cell carcinoma Parafollicular cell carcinoma Medullary carcinoma with amyloid stroma Medullary microcarcinoma	
Oxyphilic adenocarcinoma **8290**	Encapsulated angioinvasive oncocytic carcinoma of the thyroid Hurthle cell adenocarcinoma Hurthle cell carcinoma Follicular carcinoma, oxyphilic cell Minimally invasive oncocytic carcinoma of the thyroid Oncocytic adenocarcinoma Oncocytic carcinoma Widely invasive oncocytic carcinoma of the thyroid	

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Papillary thyroid carcinoma, NOS **8260** ***Note:*** For thyroid cancer only, the terms micropapillary and papillary microcarcinoma do not refer to a specific histologic type. It means that the papillary portion of the tumor is minimal or occult.	Classical (usual) papillary carcinoma Clear cell papillary thyroid carcinoma Cribriform-morular variant of PTC Hobnail variant of PTC Micropapillary thyroid carcinoma (see note) Papillary microcarcinoma (see note) Papillary thyroid carcinoma with fibromatosis/fasciitis-like stroma PTC Solid/trabecular variant of PTC Spindle cell papillary thyroid carcinoma Warthin-like papillary thyroid carcinoma	Columnar cell variant of PTC/Tall cell PTC **8344** Diffuse sclerosing PTC **8350** Encapsulated variant of PTC **8343/3** Follicular variant of papillary thyroid carcinoma **8340** Oncocytic variant of PTC **8342**
Poorly differentiated thyroid carcinoma **8337/3**	Differentiated high-grade thyroid carcinoma Insular carcinoma	

Table 13: Ovary Histologies

Table 13 list the more common histologies for ovary: **includes reportable neoplasms only**
C569 Ovary

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Coding Notes for Ovary: For ovarian primaries, code **9084/3 Teratoma with malignant transformation** when a malignant (/3) histology arises in a benign teratoma.

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma, NOS **8140**		Clear cell adenocarcinoma **8310** Endometrioid adenocarcinoma **8380** Mucinous adenocarcinoma, NOS **8480**
Adenocarcinoma of rete ovarii **9110/3**		
Adenosarcoma **8933/3**		
Adult granulosa cell tumor **8620/3**		
Carcinosarcoma, NOS **8980/3** ***Note:*** This pathology diagnosis may provide subtypes/variants of the carcinoma/adenocarcinoma component and/or sarcoma subtype/variant component.	Malignant Mixed Mullerian Tumor/MMMT ***Note***: WHO indicates this term is now a related term/synonym for carcinosarcoma	
Choriocarcinoma, NOS **9100**		
Germ cell tumor, NOS **9064**	Germinoma	Immature teratoma **9080** Dysgerminoma **9060** Yolk sac tumor, NOS **9071/3** Embryonal carcinoma **9070** Mixed germ cell tumor / mixed teratoma-yolk sac tumor **9085**
Malignant Brenner tumor **9000/3**		
Mesonephric-like adenocarcinoma **9111/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Mixed cell adenocarcinoma **8323** ***Note***: At least two histologic types must be recognized in the tumor. Percentages may be stated but do not determine histology coding. The most common mixed tumor is comprised of endometrial and clear cell carcinomas.		
Sarcoma, NOS **8800/3**		Endometrioid stromal sarcoma, high grade **8930/3** Endometrioid stromal sarcoma, low grade **8931/3** Leiomyosarcoma, NOS **8890/3** Fibrosarcoma, NOS **8810/3**
Serous carcinoma, NOS **8441**	Serous intraepithelial carcinoma **8441/2** Serous tubal intraepithelial carcinoma **8441/2** Serous endometrial intraepithelial carcinoma **8441/2** Serous cystadenocarcinoma, NOS **8441/3** Serous adenocarcinoma **8441/3** Serous papillary adenocarcinoma, NOS **8441/3** Papillary serous adenocarcinoma **8441/3** Serous surface papillary carcinoma **8441/3**	High-grade serous carcinoma/HGSC **8461/3** Low-grade serous carcinoma/micropapillary serous carcinoma **8460/3** Serous carcinoma, non-invasive, low grade **8460/2**
Small cell carcinoma hypercalcemic type **8044/3**		
Steroid cell tumor, malignant **8670/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Struma ovarii, malignant **9090/3**		
Teratoma with malignant transformation **9084/3**		
Undifferentiated carcinoma **8020/3**	Dedifferentiated carcinoma	

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Table 14: Peritoneum Histologies

Table 14 list the more common histologies for peritoneum as noted in the 5th Ed WHO Female Genital Tumors ***only***
C482 Peritoneum, NOS; peritoneal cavity

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

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

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Gastrointestinal stromal tumor **8936/3**	GIST	
High-grade serous carcinoma **8461/3**	Peritoneal serous carcinoma, high	
Low-grade serous carcinoma **8460/3**		
Mesothelioma, Malignant **9050/3**	Mesothelioma, NOS	Epithelioid mesothelioma, malignant **9052/3** Mesothelioma, biphasic **9053/3** Sarcomatoid mesothelioma **9051/3**
Sarcoma, NOS **8800/3**		Desmoplastic small round cell tumor **8806/3** Endometrioid stromal sarcoma, high-grade **8930/3** Endometrioid stromal sarcoma, low-grade **8931/3**
Solitary fibrous tumor, malignant **8815/3**		

Table 15: Fallopian Tube Histologies

Table 15 list the more common histologies for fallopian tube
C570 Fallopian tube; uterine tube

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

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

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenosarcoma **8933/3**	Mesodermal adenosarcoma	
Carcinosarcoma **8980/3** ***Note***: This pathology diagnosis may provide subtypes/variants of the carcinoma/adenocarcinoma component and/or sarcoma subtype/variant component	Malignant mixed Mullerian tumor	
Endometrioid adenocarcinoma, NOS **8380/3**		
Serous carcinoma, NOS **8441**	Serous tubal intraepithelial carcinoma (STIC) **8441/2**	High-grade serous carcinoma **8461/3** Low-grade serous carcinoma **8460/3**
Teratoma, malignant **9080/3**	Immature teratoma	

Table 16: Uterine Corpus Histologies

Table 16 list the more common histologies for uterine corpus
C540 Isthmus uteri; lower uterine segment
C541 Endometrium; endometrial gland; endometrial stroma
C542 Myometrium
C543 Fundus uteri
C548 Overlapping lesion of corpus uteri
C549 Corpus uteri; body of uterus
C559 Uterus, NOS

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenosarcoma **8933/3**	Mullerian adenosarcoma Adenocarcinoma with sarcomatous overgrowth	
Carcinoma, undifferentiated NOS **8020/3** ***Note:*** Carcinoma, undifferentiated NOS 8020/3 has been designated biologically impossible for **Myometrium (C542)** per Cancer PathCHART review.	Carcinoma, poorly differentiated Dedifferentiated carcinoma	
Carcinosarcoma, NOS **8980/3** ***Note:*** The most common carcinomas present in carcinosarcoma are endometrioid and/or serous.	Malignant mixed Mullerian tumor	
Clear cell adenocarcinoma **8310**		
Endometrioid adenocarcinoma, NOS **8380** ***Note:*** Endometrioid adenocarcinoma, NOS 8380/2 and 8380/3 have been designated biologically impossible for **Myometrium (C542)** per Cancer PathCHART review.	Endometrial adenocarcinoma/carcinoma Endometrial atypical hyperplasia/endometrioid intraepithelial neoplasia **8380/2** Mismatch repair-deficient endometrioid carcinoma **8380/3** No specific molecular profile (NSMP) endometrioid carcinoma **8380/3** P53-mutant endometrioid carcinoma **8380/3** POLE-ultramutated endometrioid carcinoma **8380/3**	Endometrioid carcinoma with squamous differentiation **8570/3**
Mesonephric adenocarcinoma **9110/3**		Mesonephric-like adenocarcinoma **9111/3**

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Mixed cell adenocarcinoma **8323** ***Note 1:*** Mixed cell adenocarcinoma is comprised of endometrial carcinoma with two distinct histological types, in which one component is either serous or clear cell. Excludes dedifferentiated carcinoma and carcinosarcoma. ***Note 2:*** Mixed cell adenocarcinoma 8323/2 or 8323/3 have been designated biologically impossible for **myometrium (C542)** per Cancer PathCHART review.		
Mucinous carcinoma, NOS **8480** ***Note:*** Mucinous carcinoma, NOS 8480/3 and 8480/3 have been designated biologically impossible for **Myometrium (C542)** per Cancer PathCHART review.		Mucinous carcinoma, intestinal type **8144**
Neuroendocrine carcinoma NOS **8246/3** ***Note***: Neuroendocrine carcinoma NOS 8246/3 has been designated biologically impossible for **Myometrium (C542)** per Cancer PathCHART review.		Large cell neuroendocrine carcinoma **8013/3** Mixed neuroendocrine non-neuroendocrine carcinoma (MiNEN) **8154/3** Small cell neuroendocrine carcinoma **8041/3**
Perivascular epithelioid tumor, malignant **8714/3**	PEComa, malignant	

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Primitive neuroendocrine tumor **9473/3**	PNET	
Sarcoma NOS **8800/3**		Endometrial stromal sarcoma, high grade **8930/3** Endometrial stromal sarcoma, low grade **8931/3** Epithelioid leiomyosarcoma **8891/3** Leiomyosarcoma NOS/spindle leiomyosarcoma **8890/3** Myxoid leiomyosarcoma **8896/3** Undifferentiated sarcoma **8805/3**
Serous carcinoma, NOS **8441**		
Squamous cell carcinoma **8070**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Table 17: Uterine Cervix Histologies

Table 17 list the more common histologies for uterine cervix
C530 Endocervix; internal os; cervical canal; endocervical canal; endocervical gland; Nabothian gland
C531 Exocervix; external os
C538 Overlapping lesion of cervix uteri; cervical stump; squamocolumnar junction of cervix
C539 Cervix uteri; cervix, NOS; uterine cervix

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

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

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

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**
Adenoid basal carcinoma **8098/3**		
Adenosarcoma **8933/3**	Adenocarcinoma with sarcomatous overgrowth	
Adenosquamous carcinoma **8560/3**		
Carcinosarcoma **8980/3** ***Note***: This pathology diagnosis may provide subtypes/variants of the carcinoma/adenocarcinoma component and/or sarcoma subtype/variant component		
Endometrioid adenocarcinoma NOS **8380/3**	Endometrial adenocarcinoma/carcinoma	
Germ cell tumor NOS **9064/3**		Choriocarcinoma NOS **9100/3** Endodermal sinus tumor/Yolk sac tumor **9071/3**
Mucoepidermoid carcinoma **8430/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Neuroendocrine carcinoma, NOS **8246/3**		Large cell neuroendocrine carcinoma **8013/3** Mixed neuroendocrine non-neuroendocrine carcinoma (MiNEN) **8154/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor, NOS **8240/3**	Neuroendocrine tumor, grade 1	Neuroendocrine tumor, grade 2 **8249/3**
Perivascular epithelioid tumor, malignant **8714/3**	PEComa, malignant	
Sarcoma, NOS **8800/3**		Endometrial stromal sarcoma, high grade **8930/3** Endometrial stromal sarcoma, low grade **8931/3** Epithelioid leiomyosarcoma **8891/3** Leiomyosarcoma NOS/spindle leiomyosarcoma **8890/3** Myxoid leiomyosarcoma **8896/3** Rhabdomyosarcoma **8900/3** Undifferentiated sarcoma **8805/3**
Squamous cell carcinoma NOS **8070/3**	SCC, NOS	Squamous cell carcinoma, HPV-associated **8085/3** Squamous cell carcinoma, HPV-independent **8086/3**

Table 18: Vagina Histologies

Table 18 list the more common histologies for vagina
C529 Vagina NOS; vaginal vault; fornix of vagina; Gartner duct; hymen

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Vagina Coding Note: p16 is a valid test to determine HPV status and can be used to code HPV associated and HPV independent histologies.

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma NOS **8140**	Adenocarcinoma, Skene, Cowper and Littre gland origin Skene/periurethral gland adenocarcinoma	Adenocarcinoma, HPV-associated **8483**
Adenoid basal carcinoma **8098**		
Adenosarcoma **8933/3**	Adenocarcinoma with sarcomatous overgrowth Mullerian adenosarcoma	
Adenosquamous carcinoma **8560**		
Carcinosarcoma **8980/3** ***Note***: This pathology diagnosis may provide subtypes/variants of the carcinoma/adenocarcinoma component and/or sarcoma subtype/variant component	Malignant mixed Mullerian tumor	
Clear cell carcinoma **8310**		
Endometrioid carcinoma **8380**	Endometrial adenocarcinoma /carcinoma	
Germ cell tumor **9064/3**		Yolk sac tumor **9071/3**
Mesonephric adenocarcinoma **9110/3**		
Mucinous carcinoma, NOS **8480**		Mucinous carcinoma, gastric type **8482** Mucinous carcinoma, intestinal type **8144**
Neuroendocrine carcinoma, NOS **8246/3**		Combined small cell neuroendocrine carcinoma **8045/3** Large cell neuroendocrine carcinoma/combined large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor, NOS **8240/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Squamous cell carcinoma NOS **8070**	SCC, NOS Squamous cell carcinoma in-situ **8070/2**	High-grade squamous intraepithelial lesion/vaginal intraepithelial neoplasia, grade 2/vaginal intraepithelial neoplasia, grade 3 **8077/2** Squamous cell carcinoma, HPV-associated **8085** Squamous cell carcinoma, HPV-independent **8086**
Undifferentiated carcinoma **8020/3**		

Table 19: Vulva Histologies

Table 19 list the more common histologies for vulva
C510 Labium majus; labia majora, NOS; Bartholin gland; Skin of labia majora
C511 Labium minus; labia minora
C512 Clitoris
C518 Overlapping lesion of vulva
C519 Vulva, NOS; external female genitalia; fourchette; labia, NOS; labium, NOS; mons pubis; mons veneris; pudendum; skin of vulva

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Vulva Coding Note: p16 is a valid test to determine HPV status and can be used to code HPV associated and HPV independent histologies.

Table begins on next page

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma **8140**	Adenocarcinoma of anogenital mammary-like glands **8140/3**	Adenocarcinoma, intestinal type **8144**
Adenoid cystic carcinoma **8200**		
Adenosquamous carcinoma **8560**		
Basal cell carcinoma **8090/3**		
Carcinoma, poorly differentiated **8020/3**		
Epithelial-myoepithelial carcinoma **8562/3**		
Germ cell tumor **9064/3**		Yolk sac tumor NOS **9071/3**
Myoepithelial carcinoma **8982/3**		
Neuroendocrine carcinoma, NOS **8246/3**		Combined small cell neuroendocrine carcinoma **8045/3** Large cell neuroendocrine carcinoma/combined large cell neuroendocrine carcinoma **8013/3** Small cell neuroendocrine carcinoma **8041/3**
Neuroendocrine tumor, NOS **8240/3**	Neuroendocrine tumor, grade 1	Neuroendocrine tumor, grade 2 **8249/3**
Paget disease, extramammary **8542/3**		
Phyllodes tumor, malignant **9020/3**		
Squamous cell carcinoma, NOS **8070**		Squamous cell carcinoma, HPV-associated **8085** Squamous cell carcinoma, HPV-independent **8086**
Sweat gland adenocarcinoma **8400**		Adenoid cystic carcinoma **8200** Apocrine adenocarcinoma **8401** Eccrine adenocarcinoma **8413** Porocarcinoma, NOS **8409**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Table 20: Soft Tissue Histologies

Table 20 list the more common histologies for soft tissue as stated in the College of American Pathologists (C.A.P.) soft tissue protocol

Note: Table 20 lists sarcomas arising in the soft tissue sites listed below only. Soft tissue neoplasms can arise in other organs. See the site-specific Solid Tumor Modules.

C490* Connective, subcutaneous and other soft tissues of head, face and neck
C491* Connective, subcutaneous and other soft tissues of upper limb and shoulder
C492* Connective, subcutaneous and other soft tissues of lower limb and leg
C493* Connective, subcutaneous and other soft tissues of thorax
C494* Connective, subcutaneous and other soft tissues of abdomen
C495* Connective, subcutaneous and other soft tissues of pelvis
C496* Connective, subcutaneous and other soft tissues of trunk
C498 Overlapping lesion of connective, subcutaneous and other soft tissues
C499* Connective, subcutaneous and other soft tissues, NOS

**For specific sites and C-codes, please refer to ICD-O-3 or ICD-O-3.1 topography lists*

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

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

- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).
- Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of histologies, including synovial sarcoma 9044/3 (column 3).

Synovial sarcoma is also a NOS because it has subtypes/variants. The subtypes/variants are indented under the NOS (synovial sarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that synovial sarcoma has subtypes/variants.

- When subtypes/variants are indented under a NOS in Column 3, use coding rules for a NOS and a single subtype/variant. For example, synovial sarcoma **9044/3** and synovial sarcoma, biphasic/synovial sarcoma, poorly differentiated **9043/3** are a NOS and a subtype/variant, **NOT** two different subtypes.

Soft Tissue Coding Notes

- This is not an exhaustive list of all malignant soft tissue tumors. If a histology is not listed, refer to the current ICD-O versions and ICD-O updates. If the term is not listed, submit your question to **Ask A SEER Registrar**.
- Soft tissue terminology used in clinical practice may differ from the terms listed in the table, ICD-O, and C.A.P. protocol. Many soft tissue histologies are compound terms and the word roots may be inverted. It is not possible to list all combinations and permutations of such compound terms. Check various permutations of the word roots in a compound term if the version is not listed in ICD-O.
  Example: Myxofibrosarcoma and fibromyxosarcoma are the same and both coded 8811/3. The word roots have been inverted.

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Angiosarcoma **9120/3**		
Epithelioid hemangioendothelioma **9133/3**	Epithelioid hemangioendothelioma with WWTR1-CAMTA1 fusion Epithelioid hemangioendothelioma with YAP1-TFE3 fusion	
Fibrosarcoma, NOS **8810/3**	Adult fibrosarcoma	Infantile fibrosarcoma **8814/3** Low-grade fibromyxoid sarcoma /Sclerosing epithelioid fibrosarcoma **8840/3** Myofibroblastic sarcoma/myofibrosarcoma **8825/3** Myxofibrosarcoma **8811/3** Solitary fibrous tumor, malignant **8815/3**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Leiomyosarcoma **8890/3**		
Liposarcoma, NOS **8850/3**		Dedifferentiated liposarcoma **8858/3** Epithelioid/Pleomorphic liposarcoma **8854/3** Myxoid liposarcoma **8852/3** Myxoid pleomorphic liposarcoma **8859/3** Well differentiated liposarcoma **8851/3**
Osteosarcoma, NOS **9180/3**	Osteosarcoma, extraskeletal	
Rhabdomyosarcoma, NOS **8900/3**		Alveolar rhabdomyosarcoma **8920/3** Ectomesenchymoma **8921/3** Embryonal rhabdomyosarcoma **8910/3** Pleomorphic rhabdomyosarcoma **8901/3** Spindle cell/sclerosing rhabdomyosarcoma **8912/3** (synonyms below) Congenital spindle cell rhabdomyosarcoma VGLL2/NCOA2/CITED2 rearrangement MYOD1-mutant spindle cell/sclerosing rhabdomyosarcoma Intraosseous spindle cell rhabdomyosarcoma (with TFCP2/NCOA2 rearrangements

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Sarcoma, NOS **8800/3** ***Note:*** Synovial Sarcoma 9040/3 is also a NOS with the following subtypes/variant: Synovial sarcoma, biphasic/synovial sarcoma, poorly differentiated 9043/3		CIC-rearranged sarcoma **9367/3** Clear cell sarcoma of soft tissue **9044/3** Epithelioid sarcoma NOS/epithelioid sarcoma classical type/epithelioid sarcoma proximal or large cell type **8804/3** Extraskeletal Ewing sarcoma **9364/3** Extraskeletal myxoid chondrosarcoma **9231/3** Mixed tumor, malignant **8940/3** Myoepithelioma, NOS/myoepithelial carcinoma **8982/3** Ossifying fibromyxoid tumor, malignant **8842/3** Phosphaturic mesenchymal tumor, malignant **8990/3** Round cell sarcoma with EWSR1-non ETS fusions **9366/3** Sarcoma with BCOR genetic alterations **9368/3** Synovial sarcoma NOS **9040/3** Synovial sarcoma, biphasic/synovial sarcoma, poorly differentiated **9043/3**
Undifferentiated sarcoma **8805/3**		Undifferentiated pleomorphic sarcoma **8802/3** Undifferentiated round cell sarcoma **8803/3** Undifferentiated spindle cell sarcoma **8801/3**

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Table 21: Bone Histologies

Table 21 list the more common histologies for bone as stated in the College of American Pathologists (C.A.P.) bone protocol
C400* Long bones of upper limbs, scapula and associated joints
C401* Short bones of upper limb and associated joints
C402* Long bones of lower limb and associated limbs
C403* Short bones of lower limb and associated joints
C408 Overlapping lesion of bones, joints and articular cartilage of limbs
C409* Bone of limb, NOS
C412* Vertebral column
C413* Rib, sternum, clavicle, and associated joints
C414* Pelvic bones, sacrum, coccyx, and associated joints
C418* Overlapping lesions of bones, joints and articular cartilage
C419* Bone, NOS

**For specific sites and C-codes, please refer to ICD-O-3 or ICD-O-3.1 topography lists*

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

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

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
- Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Bone Coding Note: This is not an exhaustive list of all malignant bone tumors. If a histology is not listed, refer to the current ICD-O versions and ICD-O updates. If the term is not listed, submit your question to **Ask A SEER Registrar**.

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adamantinoma **9261/3**	Dedifferentiated adamantinoma	
Angiosarcoma **9120/3**		
Chondrosarcoma, NOS **9220/3**	Chondrosarcoma, grade 2 Chondrosarcoma, grade 3 Fibrochondrosarcoma	Chondrosarcoma, grade 1 **9222/3** Clear cell chondrosarcoma **9242/3** Dedifferentiated chondrosarcoma **9243/3** Mesenchymal chondrosarcoma **9240/3** Periosteal chondrosarcoma **9221/3**
Chordoma, NOS **9370/3**	Poorly differentiated chordoma	Chondroid chordoma **9371/3** Dedifferentiated chordoma **9372/3**
Epithelioid hemangioendothelioma, NOS **9133/3**		
Fibrosarcoma, NOS **8810/3**		
Giant cell tumor of bone, malignant **9250/3**		
Leiomyosarcoma, NOS **8890/3**		
Osteosarcoma, NOS **9180/3**	Conventional osteosarcoma Osteoblastic sarcoma Osteogenic sarcoma, NOS Osteochondrosarcoma Osteosarcoma, extraskeletal Small cell osteosarcoma Telangiectatic osteosarcoma	High grade surface osteosarcoma **9194/3** Parosteal osteosarcoma **9192/3** Periosteal osteosarcoma **9193/3** Secondary osteosarcoma **9184/3**
Sarcoma, NOS **8800/3**		CIC-rearranged sarcoma **9367/3** Ewing sarcoma **9364/3** Round cell sarcoma with EWSR1-non ETS fusions **9366/3** Sarcoma with BCOR genetic alterations **9368/3**
Undifferentiated high grade pleomorphic sarcoma of bone **8830/3**		

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
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Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Undifferentiated pleomorphic sarcoma **8802/3**		

Table 22: Thymus Histologies

Table 22 lists the more common histologies for thymus
C379 Thymus

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Table begins on next page.

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma, NOS **8140**		Adenocarcinoma, enteric-type **8144** Low-grade papillary adenocarcinoma **8260** Thymic carcinoma with adenoid cystic carcinoma-like features **8200**
Adenosquamous carcinoma **8560***		
Carcinosarcoma **8980/3***		
Clear cell carcinoma **8310***		
Mucoepidermoid carcinoma **8430***		
Neuroendocrine carcinoma (NEC) **8246/3**		Combined small cell carcinoma **8045/3** Large cell neuroendocrine carcinoma **8013/3** Small cell carcinoma **8041/3**
Neuroendocrine tumor (NET) **8240/3**	Carcinoid tumor, NOS Neuroendocrine tumor, grade 1 Typical carcinoid	Atypical carcinoid/neuroendocrine tumor, grade 2 **8249/3**
NUT carcinoma **8023/3**		
Sarcomatoid carcinoma **8033***		
Squamous cell carcinoma, NOS **8070**		Basaloid carcinoma **8123** Basaloid squamous cell carcinoma **8083** Lymphoepithelial carcinoma **8082**
Thymic carcinoma **8586/3***	Thymoma, type C	

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Thymoma, NOS **8580** ***Note***: Subtype/variants with synonyms are separated by (/)	Intrapulmonary thymoma Metaplastic thymoma Sclerosing thymoma	Thymoma, type A/medullary/spindle cell **8581** Thymoma, type AB/mixed type **8582** Thymoma, type B1/lymphocytic/lymphocyte-rich/organoid/predominantly cortical **8583** Thymoma, type B2/cortical **8584** Thymoma, type B3/atypical/epithelial/well differentiated thymic carcinoma **8585**
Undifferentiated carcinoma **8020/3**		

*** Histologies are salivary gland-like carcinomas**

Table 23: Penis and Scrotum Histologies

Table 23 lists the more common histologies for penis
C600 Prepuce; foreskin
C601 Glans penis
C602 Body of penis; corpus cavernosum; corpus of penis
C608 Overlapping lesion of penis
C609 Penis, NOS; skin of penis
C632 Scrotum, NOS; skin of scrotum

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the **Hematopoietic Database**.

Column 1 contains specific and NOS histology terms.

- **Specific** histology terms **do not** have **subtypes/variants**
- **NOS** histology terms **do** have **subtypes/variants.**

Column 2 contains **synonyms** for the specific or NOS term. Synonyms have the **same** histology **code** as the specific or NOS term.
Column 3 contains **subtypes/variants** of the **NOS** histology. Subtypes/variants **do not** have the **same** histology code as the NOS term.
Column 3 may list more than one histology with the same ICD-O code. These histologies are separated with a slash (/).

Penis Coding Notes

- P16 is a valid test to determine HPV status and can be used to code HPV-associated and HPV-independent histologies
- When the diagnosis is SCC, HPV-associated or SCC, HPV-independent AND a subtype/variant is also stated, code the subtype variant. The subtype/variant has priority.
    - **Example**: Basaloid SCC, HPV-associated is coded basaloid SCC 8083
- Definition of HPV-associated SCC: invasive keratinizing carcinoma arising from penile mucosal or cutaneous compartments that is associated with HPV infection.
- Definition of HPV-independent SCC: invasive keratinizing carcinoma arising from penile mucosal or cutaneous compartments that is not associated with HPV infection.

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Specific and NOS Terms and Code	Synonyms	Subtypes/Variants
Adenocarcinoma, NOS **8140**		Adenosquamous carcinoma **8560** Mucoepidermoid carcinoma **8430**
Paget disease, extramammary **8542/3**		
Squamous cell carcinoma, NOS **8070** ***Note 1:*** Histologies identified with **(#)** are defined as HPV-associated neoplasms per WHO ***Note 2:*** Histologies identified with **(*)** are defined as HPV-independent neoplasms per WHO	Squamous cell carcinoma, in-situ **8070/2** SCC, NOS	Basaloid SCC **8083#** Clear cell SCC **8084#** Differentiated penile intraepithelial neoplasia **8071/2** High-grade squamous intraepithelial lesion **8077/2** Lymphoepithelial carcinoma **8082#** Papillary squamous cell carcinoma **8052*** Pseudoglandular SCC / Pseudohyperplastic **SCC 8075*** Sarcomatoid squamous cell carcinoma / spindle cell SCC **8074*** Squamous cell carcinoma, HPV-associated **8085#** Squamous cell carcinoma, HPV-independent /SCC, usual type **8086*** Verrucous carcinoma / carcinoma cuniculatum / Warty carcinoma **8051**

#: This histology is defined as HPV-associated per WHO
*: This histology is defined as HPV-independent per WHO

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Note 1: These rules are **NOT** used for tumor(s) described as metastases.
Note 2: 2007 MPH Rules and Solid Tumor rules are used based on date of diagnosis.

- Tumors diagnosed 01/01/2007 through 12/31/2022: Use the 2007 MPH Rules
- Tumors diagnosed 01/01/2023 and later: Use the Solid Tumor Rules
- The original tumor diagnosed before 1/1/2023 and a subsequent tumor diagnosed 1/1/2023 or later in the same primary site: Use the Solid Tumor Rules

Unknown if Single or Multiple Tumors

Rule M1 Abstract a **single primary**[i] when it is not possible to determine if there are **single** or **multiple** tumors.

Note 1: Use this rule only after all information sources have been exhausted.
Note 2: Examples of cases with minimal information include:

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
  - Outpatient biopsy with no follow-up information available
  - Multiple pathology reports which do not specify whether a single tumor or multiple tumors have been biopsied and/or resected

This is the end of instructions for Unknown if Single or Multiple Tumors

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

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Single Tumor

Rule M2 Abstract a **single primary**[i] when there is a **single tumor.**

Note 1: A single tumor is always a single primary
Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsite.
Note 3: The tumor may be comprised of both in situ and invasive histologies.
Note 4: The invasive malignancy may arise in or is in a background of in situ/non-invasive neoplasm.

This is the end of instructions for Single Tumors

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

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Multiple Tumors

Note 1: Multiple tumors may be single primary or multiple primaries.
Note 2: Includes combinations of in situ and invasive.

Important change to 2023 Other Sites Multiple Primary Rules: Rules M3 through M9 apply to specific sites and histologies.

Rule M3 **Acinar Adenocarcinoma (8140)** of the **prostate** is always a **single primary**[i].
Note 1: Report only one acinar/adenocarcinoma of the prostate per patient lifetime.
Note 2: 95% of prostate malignancies are the common (acinar) adenocarcinoma histology (8140/3).
Note 3: If the patient has a previous acinar adenocarcinoma of the prostate in the database and is diagnosed with adenocarcinoma in 2023, it is a single primary.
Note 4: The rule applies to multiple occurrences of acinar adenocarcinoma of prostate and/or subtype variants of acinar adenocarcinoma of prostate listed in **Table 3**.

Rule M4 Abstract **multiple primaries**[ii] when the patient has a subsequent **small cell carcinoma** of the **prostate** more than 1 year following a diagnosis of acinar adenocarcinoma and/or subtype/variant of acinar adenocarcinoma of prostate (**Table 3**).
Note 1: Small cell carcinoma (SmCC) of the prostate is rare and accounts for less than 1% of prostate cancers.
Note 2: 50% of SmCC of the prostate cases present as a de novo malignancy
Note 3: SmCC of the prostate often occurs following androgen deprivation treatment (ADVT) and/or radiation therapy for acinar adenocarcinoma
Note 4: SmCC of the prostate are aggressive with poor clinical outcomes and survival.

Rule M5 **Retinoblastoma** is always a **single primary**[i] (unilateral or bilateral).

Rule M6 **Kaposi sarcoma** (of any site(s)) is always a **single primary**[i].

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule M7 Abstract a **single primary**[i] when **follicular** and **papillary** tumors in the **thyroid** are diagnosed **within 60 days** and tumors are:

- Papillary thyroid carcinoma, NOS and follicular carcinoma, NOS **OR**
- Papillary carcinoma, follicular variant and papillary thyroid carcinoma **OR**
- Papillary carcinoma, follicular variant and follicular carcinoma **OR**
- Any papillary thyroid carcinoma subtype/variant and any follicular subtype/variant listed in Column 3, **Table 12**.

Rule M8 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are **anaplastic carcinoma** and any other histologies in the **thyroid**.

Note: This rule does not apply to multiple tumors that are anaplastic carcinoma and undifferentiated carcinoma.

Rule M9 Bilateral epithelial tumors (8000-8799) of the **ovary within 60 days** are a **single primary**[i].

Note 1: Tumors must be **same histology** or be an **NOS and subtype/variant** (are on the **same row** in **Table 13**).

Note 2: Same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3)

Rule M10 Tumors on both sides (right and left) of a site listed in **Table 1** are **multiple primaries**[ii].

Rule M11 Adenocarcinoma in **adenomatous polyposis coli** (familial polyposis) with one or more in situ or malignant polyps is a **single primary**[i].

Note: Tumors may be present in a single or multiple segments of small bowel, colon, rectosigmoid, rectum.

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule M12 Abstract **multiple primaries**[ii] when the patient has a subsequent tumor after being clinically disease-free for greater than **one year** after the original diagnosis or recurrence.

Note 1: Clinically disease-free means that there was no evidence of recurrence in the same site on follow-up.

- Scopes are NED
- Scans are NED
- All other work-up is NED

Note 2: When there is a recurrence less than or equal to one year of diagnosis, the "clock" starts over. The time interval is calculated from the date of last recurrence. In other words, the patient must have been disease-free for greater than one year from the date of the last recurrence.

Note 3: When it is unknown/not documented whether the patient had a recurrence, **use date of diagnosis** to compute the time interval.

Note 4: The physician may state this is a recurrence, meaning the patient had a previous tumor and now has another tumor. **Follow the rules**; do not attempt to interpret the physician's statement.

Rule M13 Tumors with ICD-O-3 topography codes that are different at the second (C**X**xx) and/or third characters (Cx**X**x) are **multiple primaries**[ii].

Example 1: A tumor in the penis C**6**09 and a tumor in the rectum C**2**09 have different second characters in their ICD-O-3 topography codes, so they are multiple primaries.

Example 2: A tumor in the ceric C5**3**9 and a tumor in the vulva C5**1**9 have different third characters in their ICD-O-3 topography codes, so they are multiple primaries.

Rule M14 Tumors with ICD-O-3 **topography** codes that **differ** only at the **fourth character** (Cxx**X)** and are in any **one** of the following primary sites are **multiple primaries**[ii].

- Anus and anal canal (C21_)
- Bone, joints, and articular cartilage (C40_ to C41_)
- Connective subcutaneous and other soft tissues (C49_)
- Skin (C44_)

Rule M15 A de novo (frank) in situ or malignant adenocarcinoma and an in situ or malignant tumor in a **polyp** are a **single primary**[i].

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule M16 Multiple in situ and/or malignant polyps are a **single primary**[i].
Note: Includes all combinations of adenomatous, tubular, villous, and tubulovillous adenomas or polyps.

Rule M17 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3, Table 3-23 in the Equivalent Terms and Definitions.
Note: The tumors may be subtypes/variants of the same or different NOS histologies:
- **Same NOS**: Micropapillary carcinoma of stomach 8265/3 and mucinous adenocarcinoma of stomach 8480/3 are both subtypes of adenocarcinoma, NOS of stomach but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS**: Myxofibrosarcoma 8811/3 is a subtype/variant of fibrosarcoma, NOS 8810/3; myxoid liposarcoma 8852/3 is a subtype liposarcoma, NOS 8850/3. They are distinctly different histologies. Abstract multiple primaries.

Rule M18 Abstract a **single primary**[i] when synchronous, separate/non-contiguous tumors are on **the same row in Table 3-23** in the Equivalent Terms and Definitions.
Note: The same row means the tumors are:
- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3)

Rule M19 Abstract **multiple primaries**[ii] when separate/non-contiguous tumors are on **multiple rows** in Table 2-23 in the Equivalent Terms and Definitions. Timing is irrelevant
Note: Each row in the table is a **distinctly different** histology.

Rule M20 Abstract **multiple primaries**[ii] when an **invasive** tumor occurs **more than 60 days** after an **in situ** tumor.
Note 1: This rule applies to multiple tumors, one in situ and a separate malignant tumor.
Note 2: The purpose of this rule is to ensure the case is counted as an incident (invasive) case when incidence data are analyzed.
Note 3: Abstract as multiple primaries even if the medical record/physician states it is recurrence or progression of disease.

Other Sites Multiple Primary Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule M21 Abstract a **single primary**[i] when there are multiple tumors that do not meet any of the above criteria.
Note 1: Use this rule as a last resort. Confirm that you have not overlooked an applicable rule.
Note 2: When an invasive tumor follows an in situ tumor within 60 days, abstract a single primary.

This is the end of instructions for Multiple Tumors

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

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

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.

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

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

This is a hierarchical list of source documentation.

Code the **most specific** pathology/tissue from either the resection or biopsy.
Note 1: The term "most specific" usually refers to a subtype/variant.
Note 2: The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
Note 3: When there is a discrepancy between the biopsy and resection (two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).

1. **Tissue or pathology** report from primary site (in priority order)
   A. Addendum(s) and/or comment(s)
   B. Final diagnosis/synoptic report as required by CAP
   C. CAP protocol (this is not the same as the CAP synoptic report)

Note 1: Addendums and comments on the pathology report are given highest priority because they often contain additional information about molecular testing, genetic testing, and /or special stains which give a more specific diagnosis.
Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.
Note 3: The CAP protocol is a checklist which:

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

2. **Cytology** (Fine needle biopsy from primary site, retroperitoneal, peritoneal, abdominal cavity fluid, ascites)
   Example: Fine needle aspiration of ascites shows adenocarcinoma, and the resection pathology shows serous adenocarcinoma. Code serous adenocarcinoma 8441/3

3. Tissue/pathology from metastatic site
   Note 1: Code behavior /3.
   Note 2: The **tissue** from a **metastatic** site often shows **variations** from the primary tumor. When it is the **only** tissue available, it is **more accurate** than a scan.

4. Scan: The following list is not in priority order because they are not a reliable method for identifying specific histology(ies).
   A. MRI
   B. CT
   C. PET
   D. Ultrasound

5. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following
   A. Priority order:
   B. Treatment plan
   C. Documentation from Tumor Board
   D. Documentation from the medical record that refers to the original pathology, cytology, or scan(s)
   E. Physician's **reference to** type of cancer (**histology**) in the medical record
      Note 1: Code the specific histology when documented
      Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Coding Histology

Important Information for using Other Sites Histology Tables:

- Site-specific histology tables have been added to Other Sites Solid Tumor Rules.
- Not all site groups have individual histology tables and will require the use of ICD-O and updates.
- Site-specific histology tables are based on current WHO Classification of Tumors books and the current version of ICD-O. The tables may not include all histologies that could occur in that site.
- In place of adding numerous site-based histology rules to the 2023 revision, the histology tables in Other Sites Terms and Definitions include additional coding instructions and notes to assign the correct ICD-O code when appropriate.

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

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

   Note 1: Some site-specific histologies must meet a percentage requirement in order to be coded. Refer to the Histology Rules and the appropriate site group Histology Table for coding guidance.
   Note 2: The terms above (A, B, C) must describe a **carcinoma** or **sarcoma** in order to code a histology described by those terms.
   Example: When the diagnosis is adenocarcinoma with a component of papillary **carcinoma**, code papillary carcinoma 8260.
   Negative example: When the diagnosis is simply adenocarcinoma with a papillary component. Code adenocarcinoma 8140. Do not assume this is a papillary carcinoma. This could be papillary differentiation or features.
   Note 3: When the most specific histology is described as differentiation or features, see #2.

2. Code the histology 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".
   Example: Endometrioid carcinoma with squamous differentiation has an ICD-O code of 8570/3
   Note: Do not code differentiation or features when there is no specific ICD-O code.

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

      Example: Outpatient biopsy says **consistent with adenocarcinoma**. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology to adenocarcinoma. The case meets the criteria in #**3A.**

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

      Example 1: The pathology diagnosis is adenocarcinoma consistent with tubular adenocarcinoma. The oncology consult says the patient has tubular adenocarcinoma of the stomach. This is clinical confirmation of the diagnosis, code tubular adenocarcinoma. The case meets the criteria in **bullet 1.**

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

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

List of Ambiguous Terminology

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

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

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Single Tumor: In Situ Only
(All parts are in situ)

Rule H1 Code the histology documented by the physician when the **pathology/cytology** report is **not available.**

Note 1: Priority for using documents to code histology when pathology/cytology report is not available
- Documentation in the medical record that refers to the pathologic or cytologic findings
- Physician's reference to type of cancer in the medical record

Note 2: Code the specific histology when documented.

Note 3: Code the histology to 8000/2 (cancer, in situ/non-invasive) or 8010/2 (carcinoma in situ, NOS) as stated by the physician when nothing more specific is documented.

Rule H2 Code the histology when only **one histologic type** is identified.

Note 1: Do not code terms that do not appear in the histology description.

Note 2: Use **Tables 3-23** to code histology. New codes, terms, and synonyms are included in Tables 3-23 and coding errors may occur if the table is not used.

Example: Do not code squamous cell carcinoma non-keratinizing unless the words "non-keratinizing" actually appear in the final diagnosis.

Rule H3 Code **8077/2** (squamous intraepithelial neoplasia, high grade) for the following:
- AIN, grade II/Anal intraepithelial neoplasia, grade II
- AIN, grade III/Anal intraepithelial neoplasia, grade III
- CIN with severe dysplasia
- Conjunctival intraepithelial neoplasia grade III (CIN III)
- High-grade squamous intraepithelial neoplasia, grade II
- High-grade intraepithelial neoplasia, grade III
- High-grade squamous intraepithelial lesion (HSIL)
- Intraepithelial neoplasia grade II/III
- Squamous intraepithelial neoplasia, grade II
- Squamous intraepithelial neoplasia, grade III
- Vaginal intraepithelial neoplasia, grade III/VAIN III

Note 1: Code 8077 cannot be used for glandular intraepithelial neoplasia such as pancreatic intraepithelial neoplasia (PAIN).
Note 2: This list may not include all reportable neoplasms for 8077/2. See SEER Program Coding and Staging Manual or STORE manual for reportable neoplasms.

Rule H4 Code **8148/2** (glandular intraepithelial neoplasia, grade III) for the following:

- Pancreatic intraepithelial neoplasia (PanIN III)
- High grade biliary intraepithelial neoplasms (BiIN III)
- Biliary intraepithelial neoplasm Grade 3/BiIN-3
- Esophageal intraepithelial neoplasm, high grade

Note: This list may not include all reportable neoplasms for 8148/2. See SEER Program Coding and Staging Manual or STORE manual for reportable neoplasms.

Rule H5 Code **8210** (adenocarcinoma in adenomatous polyp), **8261** (adenocarcinoma in villous adenoma), or **8263** (adenocarcinoma in tubulovillous adenoma) **only when**:

- The final diagnosis is **adenocarcinoma/carcinoma** in a polyp **OR**
- The final diagnosis is **adenocarcinoma/carcinoma**, and a residual polyp or polyp architecture is recorded in other parts of the pathology report **OR**
- The final diagnosis is **adenocarcinoma/carcinoma** and there is reference to residual or pre-existing polyp **OR**
- There is documentation that the patient had a **polypectomy**

Important note: For cases diagnosed 1/1/2023 forward: If the final diagnosis indicates a histology other than adenocarcinoma/carcinoma arising in a polyp, code the specific histology. This applies to all sites.
Example: Endometrial biopsy shows endometrioid adenocarcinoma in situ arising in a polyp. Code endometrioid adenocarcinoma, in situ.

Rule H6 Code the **subtype/variant** when a **NOS** and a **single subtype/variant** of that NOS are present.

- Adenocarcinoma in situ, NOS (**8140**) and a specific in situ adenocarcinoma
- Carcinoma in situ, NOS (**8010**) and a specific in situ carcinoma
- Melanoma in situ, NOS (**8720**) and a specific in situ melanoma
- Sarcoma, NOS (**8800**) and a specific sarcoma
- Squamous cell carcinoma, NOS (**8070**) and a specific squamous cell carcinoma

Note 1: The specific type may be identified as type, subtype, variant or predominantly.
Note 2: Do not code architecture and pattern.
Note 3: Refer to **Tables 3-23** in Terms and Definitions for additional coding instructions. There may be exceptions to this rule.

Rule H7 Code a **combination code** when there are multiple specific in situ histologies or when there is an NOS with multiple specific in situ histologies **AND**

- The combination is listed in **Table 2** in Equivalent Terms and Definitions, ICD-O and all updates **OR**
- You receive a combination code from **Ask A SEER Registrar**

Note 1: The rules are hierarchical. Use this rule when previous rules do not apply.
Note 2: Submit a question to Ask A SEER Registrar when a combination is not listed in Table 2 in Equivalent Terms and Definitions, ICD-O, and all ICD-O updates.

This is the end of instructions for a Single Tumor: In Situ Components
Code the histology according to the rule that fits the case

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 3-23**, 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 3-23**, ICD-O, and ICD-O updates, ignore the in situ term.

This is the end of instructions for a Single Tumor: Invasive and In Situ Components
Code the histology according to the rule that fits the case

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Single Tumor: Invasive Only

Rule H9 Code the histology documented by the physician when the **pathology/cytology** report is **not available**.

Note 1: Priority for using documents to code histology when pathology/cytology report is not available

- Documentation in the medical record that refers to the pathologic or cytologic findings
- Physician's reference to type of cancer in the medical record
- CT, PET, or MRI scans

Note 2: Code the specific histology when documented.

Note 3: Code the histology to 8000/3 (cancer, malignant neoplasm) or 8010/3 (carcinoma, NOS) as stated by the physician when nothing more specific is documented.

Rule H10 Code the histology from a **metastatic site** when there is **no pathology/cytology** from the primary site.

Note: Code the behavior /3.

Rule H11 Code **8140** (adenocarcinoma, NOS) for **prostate primaries** when the diagnosis is:

- Acinar adenocarcinoma/carcinoma **OR**
- Adenocarcinoma **OR**
- Adenocarcinoma with ductal features **OR**
- Atrophic adenocarcinoma **OR**
- Foamy gland adenocarcinoma **OR**
- Microcystic adenocarcinoma **OR**
- Pseudohyperplastic adenocarcinoma **OR**
- Prostatic intraepithelial-like carcinoma

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

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 3-23, 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.

Rule H13 Code **8210** (adenocarcinoma in adenomatous polyp), **8261** (adenocarcinoma in villous adenoma), or **8263** (adenocarcinoma in tubulovillous adenoma) **only when:**

- The final diagnosis is **adenocarcinoma/carcinoma** in a polyp **OR**
- The final diagnosis is **adenocarcinoma/carcinoma**, and a residual polyp or polyp architecture is recorded in other parts of the pathology report **OR**
- The final diagnosis is **adenocarcinoma/carcinoma** and there is reference to residual or pre-existing polyp **OR**
- There is documentation that the patient had a polypectomy

Important note for cases diagnosed 1/1/2023 forward: If the final diagnosis indicates a histology other than adenocarcinoma/carcinoma arising in a polyp, code the specific histology.

Example: Cervix biopsy shows endometrioid adenocarcinoma arising in multiple polyps. Code endometrioid adenocarcinoma.

Rule H14 Code the subtype/variant for pancreas primaries when the diagnosis is **ductal carcinoma/adenocarcinoma AND**

- Adenosquamous carcinoma **8560/3**
- Colloid/mucinous carcinoma/adenocarcinoma **8480/3**
- Hepatoid carcinoma **8576/3**
- Large cell carcinoma with rhabdoid phenotype **8014/3**
- Medullary carcinoma **8510/3**
- Signet-ring/poorly cohesive carcinoma/adenocarcinoma **8490/3**
- Undifferentiated carcinoma **8020/3**
- Undifferentiated carcinoma with osteo-clast-like giant cells **8035/3**

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule H15 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS, such as the following:

- Cancer/malignant neoplasm, NOS (**8000**) **AND** a subtype/variant of cancer
- Carcinoma, NOS (**8010**) **AND** a subtype/variant of carcinoma
- Adenocarcinoma, NOS (**8140**) **AND** a subtype/variant of adenocarcinoma
- Squamous cell carcinoma, NOS (**8070**) **AND** a subtype/variant of adenocarcinoma
- Melanoma, NOS (**8720**) **AND** a subtype/variant of melanoma
- Sarcoma, NOS (**8800**) **AND** a subtype/variant of sarcoma

Note: See **Tables 3-23** to find NOS and subtype/variants. There may be exceptions to this rule.

Rule H16 Code anaplastic carcinoma of thyroid (**8021**) or undifferentiated carcinoma of thyroid (**8020**) when other thyroid histologies are present in a **single** tumor.

- Treatment and prognosis will be largely determined by the anaplastic or undifferentiated component.
- This rule is new for 2023

Rule H17 Code **dedifferentiated carcinoma** (**8020**) when mixed with endometrioid carcinoma/adenocarcinoma.

- Dedifferentiated carcinoma is a distinct entity which has worse prognosis than endometrioid adenocarcinoma.

Rule H18 Code **papillary carcinoma/adenocarcinoma** of the **thyroid** to papillary adenocarcinoma, NOS (**8260**).

Rule H19 Code **papillary microcarcinoma** of **thyroid** to papillary adenocarcinoma, NOS (**8260**).

Note: For thyroid primaries only, the term micropapillary/papillary microcarcinoma does not refer to a specific histologic type. In North America, it means the papillary component of the tumor is minimal or occult.

Rule H20 Code **papillary carcinoma, follicular variant** of **thyroid (8340)** when there are multiple papillary and follicular carcinoma subtypes/variants:

- Papillary thyroid carcinoma, NOS and follicular carcinoma, NOS **OR**
- Papillary carcinoma, follicular variant and papillary thyroid carcinoma **OR**
- Papillary carcinoma, follicular variant and follicular carcinoma **OR**
- Any papillary thyroid carcinoma subtype/variant and any follicular subtype/variant listed in Column 3, **Table 12**

Note: Some thyroid histologies are compound terms meaning two or more histology types are combined into a single ICD-O code. Use **Table 12**, ICD-O, and all ICD-O updates to determine if the term containing multiple histologies has a specific code.

Rule H21 Code a combination code when there are multiple specific histologies or when there is an NOS with multiple specific histologies **AND**

- The combination is listed in **Table 2** in Equivalent Terms and Definitions, ICD-O and all updates **OR**
- There are coding instructions for the combination in the applicable histology **Tables 3-23** **OR**
- You receive a combination code from **Ask A SEER Registrar**

Note 1: The rules are hierarchical. Use this rule when previous rules do not apply.

Note 2: Submit a question to **Ask A SEER Registrar** when a combination is not listed in Table 2 in Equivalent Terms and Definitions, ICD-O, and all ICD-O updates.

This is the end of instructions for a Single Tumor: Invasive Only
Code the histology according to the rule that fits the case

Multiple Tumors Abstracted as a Single Primary

Rule H22 Code the histology documented by the physician when the **pathology/cytology** report is **not available.**

Note 1: Priority for using documents to code histology when pathology/cytology report is not available

- Documentation in the medical record that refers to the pathologic or cytologic findings
- Physician's reference to type of cancer in the medical record
- CT, PET, or MRI scans

Note 2: Code the specific histology when documented.

Note 3: Code the histology to 8000/3 (cancer, malignant neoplasm) or 8010/3 (carcinoma, NOS) as stated by the physician when nothing more specific is documented.

Rule H23 Code the histology from a **metastatic site** when there is **no pathology/cytology** from the primary site.

Note: Code the behavior /3.

Rule H24 Code **8140** (adenocarcinoma, NOS) for **prostate primaries** when the diagnosis is:

- Acinar adenocarcinoma/carcinoma **OR**
- Adenocarcinoma **OR**
- Adenocarcinoma with ductal features **OR**
- Atrophic adenocarcinoma **OR**
- Foamy gland adenocarcinoma **OR**
- Microcystic adenocarcinoma **OR**
- Pseudohyperplastic adenocarcinoma **OR**
- Prostatic intraepithelial-like carcinoma

Rule H25 Code **8077/2** (Squamous intraepithelial neoplasia, high grade) for the following:

- AIN, grade II/Anal intraepithelial neoplasia, grade II
- AIN, grade III/Anal intraepithelial neoplasia, grade III
- CIN with severe dysplasia
- Conjunctival intraepithelial neoplasia grade III (CIN III)
- High-grade squamous intraepithelial neoplasia, grade II
- High-grade intraepithelial neoplasia, grade III
- High-grade squamous intraepithelial lesion (HSIL)
- Intraepithelial neoplasia grade II/III
- Squamous intraepithelial neoplasia, grade II
- Squamous intraepithelial neoplasia, grade III
- Vaginal intraepithelial neoplasia, grade III/VAIN III

Note 1: Code 8077 cannot be used for glandular intraepithelial neoplasia such as pancreatic intraepithelial neoplasia (PAIN).
Note 2: This list may not include all reportable neoplasms for 8077/2. See SEER Program Coding and Staging Manual or STORE manual for reportable neoplasms.

Rule H26 Code **8148/2** (Glandular intraepithelial neoplasia grade III) for the following:

- Pancreatic intraepithelial neoplasia (PanIN III)
- High grade biliary intraepithelial neoplasms (BiIN III)
- Biliary intraepithelial neoplasm Grade 3/BiIN-3
- Esophageal intraepithelial neoplasm, high grade

Note: This list may not include all reportable neoplasms for 8148/2. See SEER Program Coding and Staging Manual or STORE manual for reportable neoplasms.

Rule H27 Code the histology when only **one histologic type** is identified.
Note: 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

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule H28 Code the histology of the underlying tumor when there is **extramammary Paget disease** and an underlying tumor of the **anus, perianal region, or vulva**.

Rule H29 Code **8210** (adenocarcinoma in adenomatous polyp), **8261** (adenocarcinoma in villous adenoma), or **8263** (adenocarcinoma in tubulovillous adenoca) **only when**:

- The final diagnosis is **adenocarcinoma/carcinoma** in a polyp **OR**
- The final diagnosis is **adenocarcinoma/carcinoma**, and a residual polyp or polyp architecture is recorded in other parts of the pathology report **OR**
- The final diagnosis is **adenocarcinoma/carcinoma** and there is reference to residual or pre-existing polyp **OR**
- There is documentation that the patient had a polypectomy

Important note for cases diagnosed 1/1/2023 forward: If the final diagnosis indicates a histology other than adenocarcinoma/carcinoma arising in a polyp, code the specific histology.

Example: Cervix biopsy shows endometrioid adenocarcinoma arising in multiple polyps. Code endometrioid adenocarcinoma.

Rule H30 Code **papillary carcinoma, follicular variant** of **thyroid** (**8340**) when there are multiple papillary and follicular carcinoma subtypes/variants:

- Papillary thyroid carcinoma, NOS and follicular carcinoma, NOS **OR**
- Papillary carcinoma, follicular variant and papillary thyroid carcinoma **OR**
- Papillary carcinoma, follicular variant and follicular carcinoma **OR**
- Any papillary thyroid carcinoma subtype/variant and any follicular subtype/variant listed in Column 3, **Table 12**

Rule H31 Code **papillary microcarcinoma** of thyroid to papillary carcinoma/adenocarcinoma of the **thyroid** to **8260.**

Note: For thyroid primaries only, the term micropapillary/papillary microcarcinoma does not refer to a specific histologic type. In North America, it means the papillary component of the tumor is minimal or occult.

Rule H32 Code the single **invasive** histology for **combinations of invasive and in situ**. Ignore the in situ terms.

Note: If the Multiple Primary Rules indicate an invasive tumor and separate in situ tumor are a single primary, code the invasive histology.

Other Sites Histology Rules
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia
For Cases Diagnosed 1/1/2023 Forward

Rule H33 Code the subtype/variant for **pancreas** primaries when the diagnosis is **ductal carcinoma/adenocarcinoma** **AND**

- Adenosquamous carcinoma **8560/3**
- Colloid/mucinous carcinoma/adenocarcinoma **8480/3**
- Hepatoid carcinoma **8576/3**
- Large cell carcinoma with rhabdoid phenotype **8014/3**
- Medullary carcinoma **8510/3**
- Signet-ring/poorly cohesive carcinoma/adenocarcinoma **8490/3**
- Undifferentiated carcinoma **8020/3**
- Undifferentiated carcinoma with osteo-clast-like giant cells **8035/3**

Rule H34 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS, such as the following:

- Cancer/malignant neoplasm, NOS (**8000**) **AND** a subtype/variant of cancer
- Carcinoma, NOS (**8010**) **AND** a subtype/variant of carcinoma
- Adenocarcinoma, NOS (**8140**) **AND** a subtype/variant of adenocarcinoma
- Squamous cell carcinoma, NOS (**8070**) **AND** a subtype/variant of adenocarcinoma
- Melanoma, NOS (**8720**) **AND** a subtype/variant of melanoma
- Sarcoma, NOS (**8800**) **AND** a subtype/variant of sarcoma

Note: See **Tables 3-23** in to find NOS and subtype/variants. There may be exceptions to this rule.

Rule H35 Code a combination code when there are multiple specific histologies or when there is an NOS with multiple specific histologies **AND**

- The combination is listed in **Table 2** in Equivalent Terms and Definitions, ICD-O and all updates OR
- There are coding instructions for the combination in the applicable histology **Tables 3-23** OR
- You receive a combination code from Ask A SEER Registrar

Note 1: The rules are hierarchical. Use this rule when previous rules do not apply.
Note 2: Submit a question to **Ask A SEER Registrar** when a combination is not listed in Table 2 in Equivalent Terms and Definitions, ICD-O, and all ICD-O updates.

This is the end of instructions for Multiple Tumors Abstracted as a Single Primary
Code the histology according to the rule that fits the case