# Solid Tumor Rules

Effective with Cases Diagnosed 1/1/2018 and Forward

**Updated September 2021**


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Suggested citation: Dickie, L., Johnson, CH., Adams, S., Negoita, S. (September 2021). Solid Tumor Rules. National Cancer Institute, Rockville, MD 20850.

# Solid Tumor Rules
# Preface
# (Excludes lymphoma and leukemia M9590 – M9992)

## 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 is planned to be updated for 2023 (the 2007 MPH Other Sites applies to 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 2018**
**General Instructions**
**(Excludes lymphoma and leukemia M9590 – M9992)**

## 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 rules
    - Other Sites rules will be revised for 2023 implementation. The Solid Tumor Task Force has identified the need to expand the rules to include GYN, soft tissue, thyroid as well as other site-specific solid tumors

**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 2018 Solid Tumor Rules) AND **always include primary site and diagnosis year.**

## 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
- 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

**Solid Tumor Rules 2018**
**General Instructions**
**(Excludes lymphoma and leukemia M9590 – M9992)**

## 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 2018**
**General Instructions**
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## 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-M9992.

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:
    - Other Sites (not updated for 2018) for solid tumors which occur in primary sites not covered by the site-specific rules.
6. 2007 MPH Rules. 2018 Solid Tumor Rules, and 2021 Cutaneous Melanoma rules are used based on **date of diagnosis**. See the site-specific rules for instruction on which rules to use.
    - 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 (with exceptions in #4)
    - 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 2018 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
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

## 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**).

## 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.

## 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.

- **Clinically disease-free** means that there was **no <u>evidence</u>** of recurrence on follow-up.
- 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".
- No evidence of disease (NED) means complete response to treatment.

## 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) have released the 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, and 2022 Guidelines for ICD-O-3.2 Histology Code and Behavior Update, effective for cases diagnosed 1/1/2022 forward. The updates include:

- New ICD-O codes
- Changes in behaviors for existing ICD-O codes
- New preferred terminology

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 which can be found at: https://seer.cancer.gov/icd-o-3/
- 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 2018**
**General Instructions**
**(Excludes lymphoma and leukemia M9590 – M9992)**

## 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 (M9590-9992).**

**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)

**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".

**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 – M9992 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.

## Changes from 2007 MPH Rules

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

1. **NST (No Special Type), 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 longer** 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".

## 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

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

- 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**

## 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.**

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Terms and Descriptive Language | Site Term and Code |
|---|---|
| Areolar<br>Nipple<br>Paget disease **without** underlying tumor<br>***Note:*** Paget with underlying tumor is coded to the quadrant of breast in which the underlying tumor is located | Nipple **C500** |
| Above nipple<br>Area extending 1 cm around areolar complex<br>Behind the nipple<br>Below the nipple<br>Beneath the nipple<br>Central portion of breast<br>Cephalad to nipple<br>Infra-areolar<br>Lower central<br>Next to areola NOS<br>Next to nipple<br>Retroareolar<br>Subareolar<br>Under the nipple<br>Underneath the nipple | Central portion of breast **C501** |
| Superior inner<br>Superior medial<br>Upper inner quadrant (UIQ)<br>Upper medial | Upper inner quadrant of breast **C502** |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Terms and Descriptive Language | Site Term and Code |
|---|---|
| Inferior inner<br>Inferior medial<br>Lower inner quadrant (LIQ)<br>Lower medial | Lower inner quadrant of breast **C503** |
| Superior lateral<br>Superior outer<br>Upper lateral<br>Upper outer quadrant (UOQ) | Upper outer quadrant of breast **C504** |
| Inferior lateral<br>Inferior outer<br>Lower lateral<br>Lower outer quadrant (LOQ) | Lower outer quadrant of breast **C505** |
| Axillary tail of breast<br>Tail of breast NOS<br>Tail of Spence | Axillary tail of breast **C506** |
| 12:00 o'clock<br>3:00 o'clock<br>6:00 o'clock<br>9:00 o'clock<br>Inferior breast NOS<br>Inner breast NOS<br>Lateral breast NOS<br>Lower breast NOS<br>Medial breast NOS<br>Midline breast NOS<br>Outer breast NOS<br>Overlapping lesion of breast<br>Superior breast NOS<br>Upper breast NOS | Overlapping lesion of breast C**508**<br><br>***Note:*** This is a **single tumor** which **overlaps quadrants/subsite.** |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Terms and Descriptive Language | Site Term and Code |
|---|---|
| ¾ or more of breast involved with tumor<br>Diffuse (tumor size 998)<br>Entire breast<br>Inflammatory without palpable mass<br>Multiple tumors in different subsites (quadrants) within the same breast | Breast NOS **C509**<br><br>***Note:*** Used for:<br>• Non-contiguous **multiple** tumors in **different quadrants/subsites** of same breast **OR**<br>• **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.)<br>• Inflammatory carcinoma; diffuse tumor |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 and Kaposi sarcoma M9140)**

| Required Histology Terms | Histology Combination Term and Code |
|---|---|
| DCIS/duct carcinoma/carcinoma NST **8500**<br><br>**AND**<br><br>Lobular carcinoma **8520**<br><br>***Note 1:*** Both histologies, duct and lobular, **must have** the **same behavior** code.<br>***Note 2:*** **8522** is used when:<br>• Duct **AND** lobular carcinoma are present in a **single tumor** **OR**<br>• Duct is present in at least one tumor and lobular is present in at least one tumor in the same breast **OR**<br>• One tumor is mixed duct and lobular; the other tumor in the same breast is either duct or lobular **OR**<br>• All tumors in the same breast are mixed duct and lobular<br>***Example***: One tumor with invasive duct CA in LOQ RT breast; second tumor with invasive lobular in UOQ RT breast<br>***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.** | **Invasive** carcinoma NST/duct carcinoma **and invasive** lobular carcinoma **8522/3**<br>***Note 1*:** CAP uses the term Invasive carcinoma with ductal and lobular features ("mixed type carcinoma")<br>***Note 2:*** This is the exception to the instruction that features are not coded.<br>***Note 3:*** Carcinoma NST includes all subtypes/variants of carcinoma NST.<br><br>DCIS **and in situ** lobular carcinoma **8522/2**<br>***Note:*** The lobular carcinoma includes pleomorphic lobular carcinoma in situ **8519/2.** |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 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<br><br>**AND**<br><br>**Any** histology in **Table 3 with exception** of<br>• Lobular carcinoma **8520** and pleomorphic lobular carcinoma in situ **8519/2***<br>• Paget disease **8540**<br><br>***Note 1:*** Both histologies **must have** the **same behavior** code.<br>***Note 2:*** See **Table 3** for carcinoma NST/duct carcinoma subtypes/variants.<br>***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**<br><br>DCIS mixed with other in situ carcinoma **8500/2**<br>***Note***: Prior to 2018, DCIS and other in situ was coded **8523/2**. |
| **Lobular** carcinoma<br><br>**AND**<br><br>**Any** histology in **Table 3 with exception** of<br>• Duct carcinoma/carcinoma NST/DCIS (and subtypes/variants) **8500**<br>• Paget disease, in situ and invasive<br><br>***Note 1:*** See **Table 3** for carcinoma NST/duct carcinoma subtypes/variants.<br>***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**<br><br>In situ lobular mixed with other types of in situ carcinoma **8524/2** |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Required Histology Terms | Histology Combination Term and Code |
|---|---|
| **Metaplastic carcinoma OR any ONE subtype/variant of metaplastic carcinoma**<br><br>**AND**<br><br>**Duct carcinoma/carcinoma NST OR**<br><br>**Lobular carcinoma** | Code metaplastic carcinoma **8575** OR<br>Subtype/variant of metaplastic carcinoma<br><br>**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**<br><br>**AND**<br><br>**Underlying** DCIS<br><br>***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**<br><br>**Paget disease** (specified as in situ) and DCIS/intraductal carcinoma **8543/2** |
| **Paget** disease<br><br>**AND**<br><br>**Underlying** infiltrating **duct** carcinoma/carcinoma NST and **all subtypes/variants** of infiltrating duct/carcinoma NST (must be a /3)<br><br>***Note:*** See **Table 3** for subtypes/variants of carcinoma NST/duct carcinoma. | Paget disease and infiltrating duct carcinoma **8541/3** |
| **Any** two invasive carcinoma NST subtypes/variants (percentage not stated) abstracted as a single primary<br>***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.<br>***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 – M9992 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 – M9992 and Kaposi sarcoma M9140)**

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

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific and NOS/NST Terms and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| | DCIS of intermediate nuclear grade 8500/2<br>DCIS of low nuclear grade 8500/2<br>Duct/ductal carcinoma<br>Duct/ductal carcinoma in situ **8500/2**<br>Duct/ductal carcinoma NOS<br>Duct/ductal carcinoma NST (no special type)<br>Duct/ductal carcinoma with apocrine features<br>Duct/ductal carcinoma with apocrine metaplasia<br>Duct/ductal carcinoma with lobular features<br>Duct/ductal carcinoma with micropapillary features<br>Duct/ductal carcinoma with mucin production<br>Duct/ductal carcinoma with neuroendocrine features<br>Duct/ductal carcinoma with squamous metaplasia Infiltrating ductal carcinoma **8500/3**<br>Intraductal carcinoma **8500/2**<br>Invasive carcinoma with medullary features 8500/3<br>Invasive carcinoma with micropapillary features **8500/3** | |

| Specific and NOS/NST Terms and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| | Invasive carcinoma with neuroendocrine features **8500/3**<br>Invasive carcinoma not otherwise specified (ductal/NOS) **8500/3**<br>Invasive carcinoma NST with metaplastic features **8500/3**<br>Invasive carcinoma NST/duct with medullary features **8500/3**<br>Invasive carcinoma, with signet-ring cell features **8500/3**<br>Invasive carcinoma of no special type (NST) **8500/3**<br>Invasive carcinoma with clear cell (glycogen rich) features **8500/3**<br>Invasive carcinoma, NST **8500/3**<br>Invasive carcinoma, type cannot be determined **8500/3**<br>Invasive mammary carcinoma **8500/3**<br>Invasive mammary carcinoma associated with encysted papillary carcinoma **8500/3**<br>Invasive mammary carcinoma NST with lobular features **8500/3**<br>Invasive mammary carcinoma NST with medullary features **8500/3**<br>Invasive mammary carcinoma NST with mucinous features **8500/3**<br>Invasive mammary carcinoma NST with neuroendocrine features **8500/3** | |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific and NOS/NST Terms and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| | Invasive mammary carcinoma NST with tubulo-lobular variant **8500/3**<br>Invasive mammary carcinoma with apocrine features **8500/3**<br>Invasive mammary carcinoma with cribriform features **8500/3**<br>Invasive mammary carcinoma with tubular features **8500/3**<br>Mammary carcinoma in situ **8500/2**<br>Mammary carcinoma/cancer<br>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<br>Classic lobular carcinoma<br>Florid lobular carcinoma **8520/2**<br>Intraductal papilloma with lobular carcinoma in situ **8520/2**<br>Invasive lobular carcinoma, alveolar type/variant **8520/3**<br>Invasive lobular carcinoma, solid type **8520/3**<br>Lobular carcinoma in situ **8520/2**<br>Lobular carcinoma with cribriform features | Pleomorphic lobular carcinoma in situ **8519/2***<br><br>***Note:*** 8519/2 is a new code for in situ /2 tumors only. |

| Specific and NOS/NST Terms and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| | Mixed lobular carcinoma (lobular carcinoma NOS and one or more variants of lobular carcinoma)<br>Invasive pleomorphic lobular carcinoma **8520/3**<br>Solid lobular carcinoma<br>Tubulolobular carcinoma | |
| **Medullary carcinoma 8510** | MC | Medullary carcinoma with lymphoid stroma **8512**<br>Atypical medullary carcinoma (AMC) **8513** |
| **Metaplastic carcinoma NOS or of no special type (NST) 8575**<br><br>***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.<br><br>***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<br>Metaplastic carcinoma, mixed epithelial and mesenchymal type<br>Metaplastic carcinoma with mesenchymal differentiation<br>Metaplastic carcinoma with squamous features<br>Metaplastic carcinoma with other types of mesenchymal differentiation<br>Mixed metaplastic carcinoma | Carcinosarcoma **8980/3**<br>Fibromatosis-like metaplastic carcinoma **8572**<br>Low grade adenosquamous carcinoma **8560**<br>Metaplastic carcinoma spindle-cell type/spindle cell carcinoma **8032**<br>Metaplastic carcinoma with chondroid differentiation/with osseous differentiation **8571**<br>Myoepithelial carcinoma **8982**<br>Sarcomatoid carcinoma **8033**<br>Squamous cell carcinoma **8070** |
| **Mucinous carcinoma 8480** | Colloid carcinoma<br>Mucinous adenocarcinoma | |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

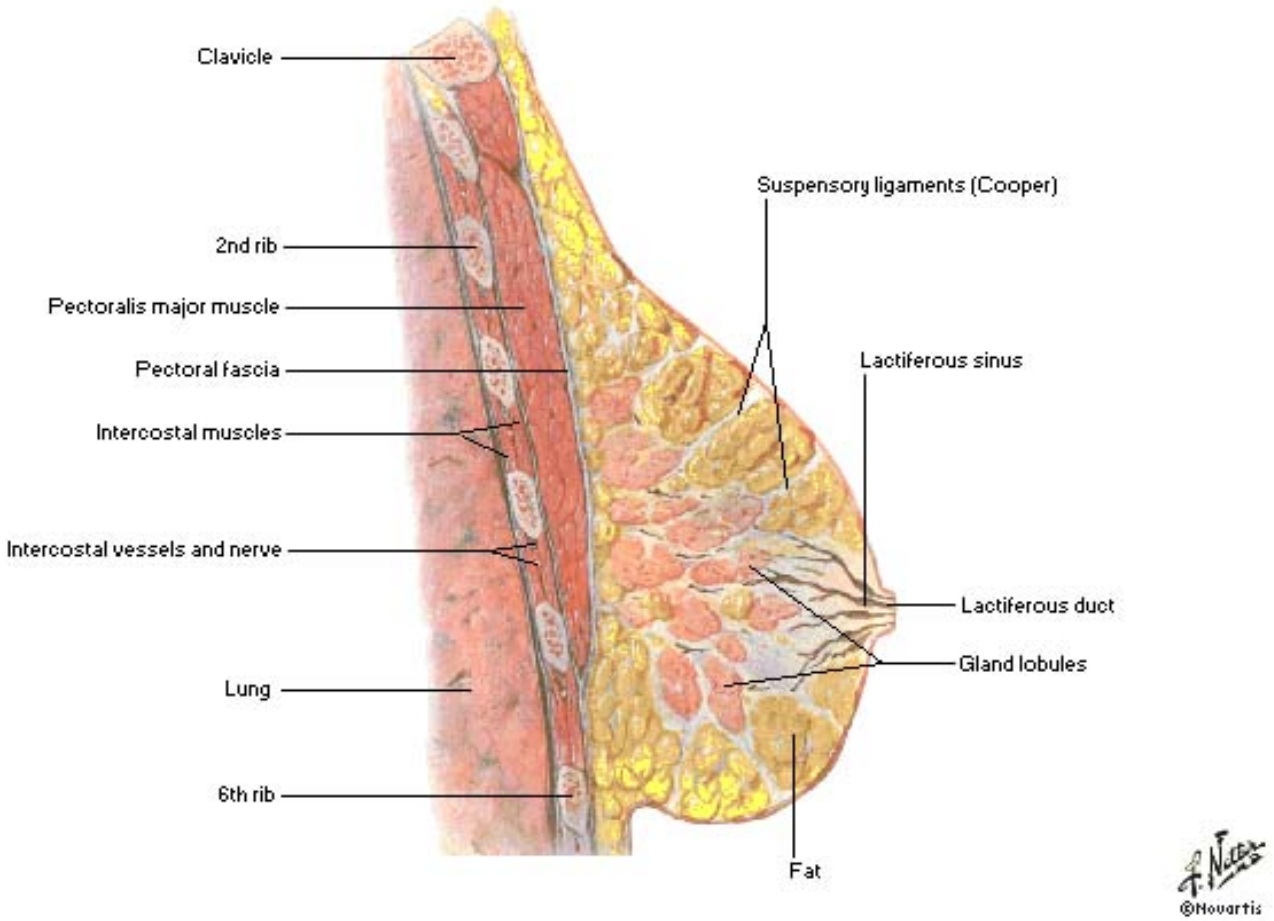
| Specific and NOS/NST Terms and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| *Note 1:* This is a diagnosis that is **EXACTLY** "mucinous carcinoma," "mucinous duct carcinoma," "mucinous DCIS" OR "greater than 90% mucinous." **See Histology Rules**.<br>*Note 2:* Mucinous duct carcinoma is listed on the CAP protocol. It is not recognized by WHO or IARC. Mucinous carcinoma is not a subtype/variant of Carcinoma NST/duct carcinoma. | 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*** <br>Intraductal papillary carcinoma with DCIS **8503/2*** <br>Intraductal papilloma with ductal carcinoma in situ **8503/2** <br>Invasive ductal papillary carcinoma **8503/3** <br>Invasive papillary carcinoma **8503/3** <br>Papillary carcinoma of breast, NOS **8503/3** <br>Papillary carcinoma non-invasive **8503/2*** <br>Papillary ductal carcinoma in situ **8503/2*** | Encapsulated papillary carcinoma **8504** <br>non-infiltrating/intracystic **8504/2** <br>with invasion **8504/3** <br>with invasive carcinoma, NST/invasive duct carcinoma **8504/3** <br>Micropapillary carcinoma **8507*** <br>Tall cell carcinoma with reverse polarity **8509/3** <br>Solid papillary carcinoma in situ **8509/2*** <br>with invasion **8509/3*** |

**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

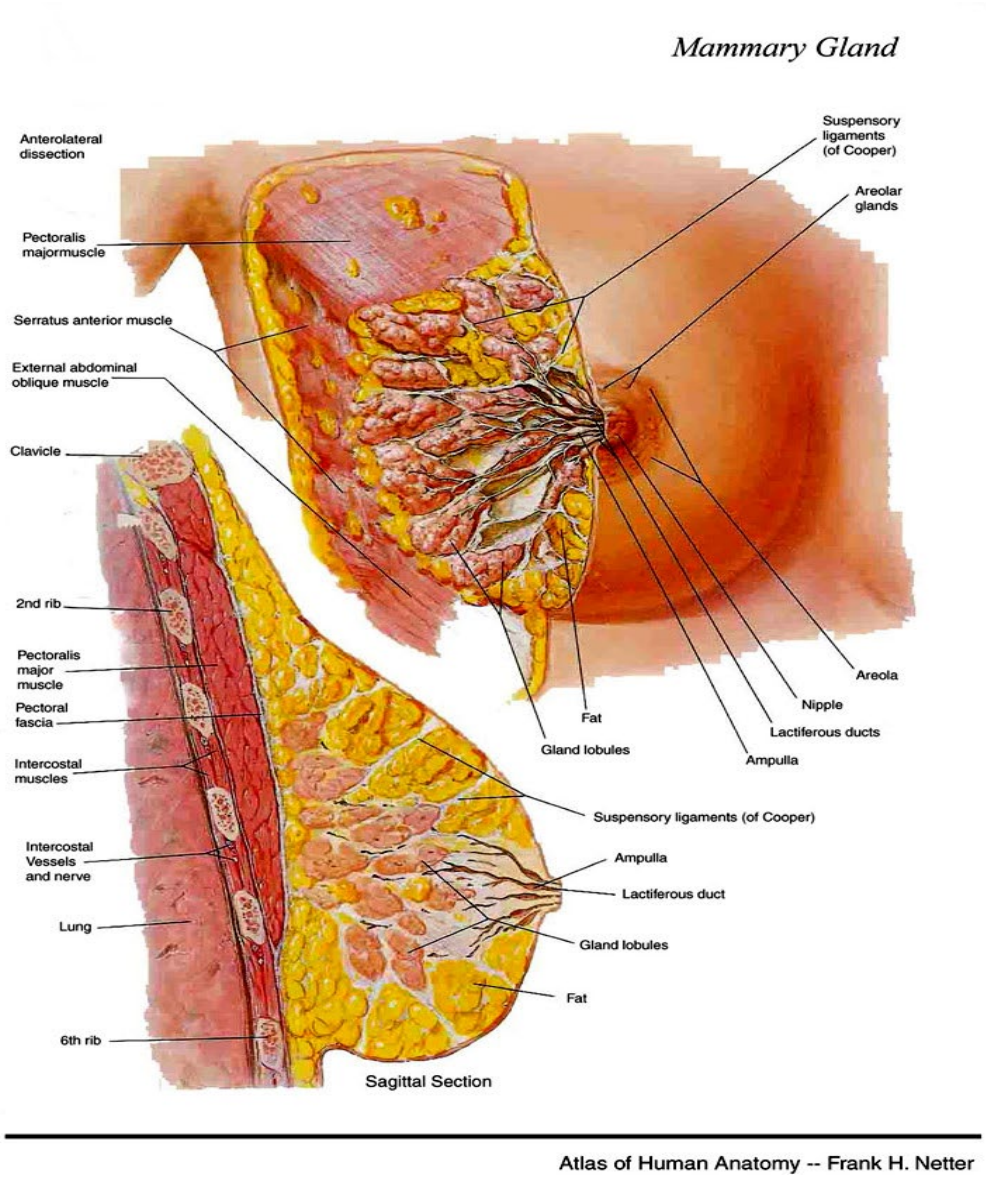
| Specific and NOS/NST Terms and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Phyllodes tumor, malignant 9020/3** | Cystosarcoma phyllodes, malignant<br>Periductal stromal tumor, low grade | |
| **Polymorphous carcinoma 8525** | | |
| **Sarcoma NOS 8800/3**<br><br>*Note 1:* Angiosarcoma **9120/3** is also a NOS with the following subtypes/variants:<br>Lymphangiosarcoma 9170/3<br>Malignant hemangioendothelioma 9130/3<br><br>*Note 2*: Rhabdomyosarcoma **8900/3** is also a NOS with the following subtypes/variants:<br>Alveolar type rhabdomyosarcoma **8920/3**<br>Embryonal type rhabdomyosarcoma **8910/3**<br>Pleomorphic rhabdomyosarcoma **8901/3** | | Angiosarcoma **9120/3**<br>Epithelioid angiosarcoma<br>Hemangiosarcoma<br>Lymphangiosarcoma **9170/3**<br>Malignant hemangioendothelioma **9130/3**<br>Liposarcoma **8850/3**<br>Leiomyosarcoma **8890/3**<br>Osteosarcoma **9180/3**<br>Rhabdomyosarcoma **8900/3**<br>Alveolar type **8920/3**<br>Embryonal type **8910/3**<br>Pleomorphic **8901/3** |
| **Sebaceous carcinoma 8410** | | |
| **Secretory carcinoma 8502** | Juvenile breast carcinoma | |
| **Signet ring carcinoma 8490** | | |
| **Small cell carcinoma 8041** | Carcinoid tumor of breast<br>Endocrine carcinoma<br>Neuroendocrine carcinoma, poorly differentiated | Carcinoma with neuroendocrine differentiation **8574/3**<br>Neuroendocrine tumor, well-differentiated **8246/3** |
| **Tubular carcinoma 8211** | | |

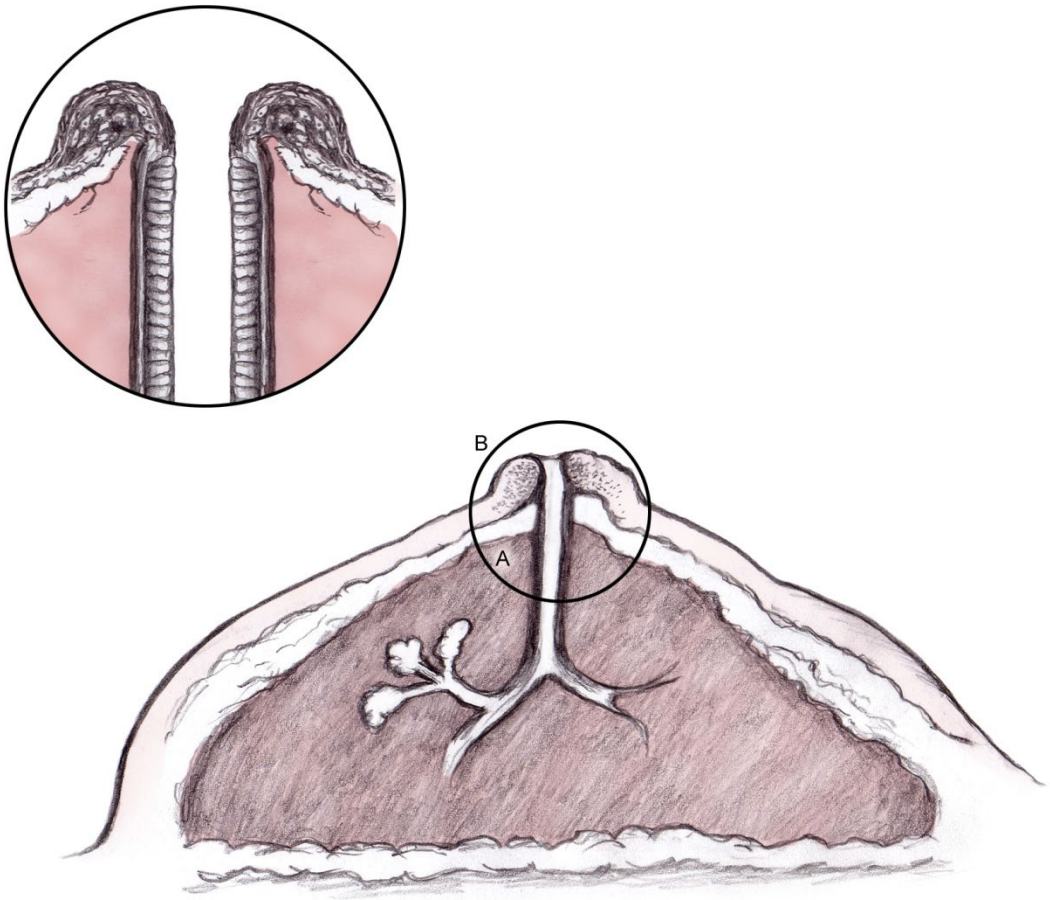
***New codes approved by IARC/WHO Committee for ICD-O**

## Illustrations



**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

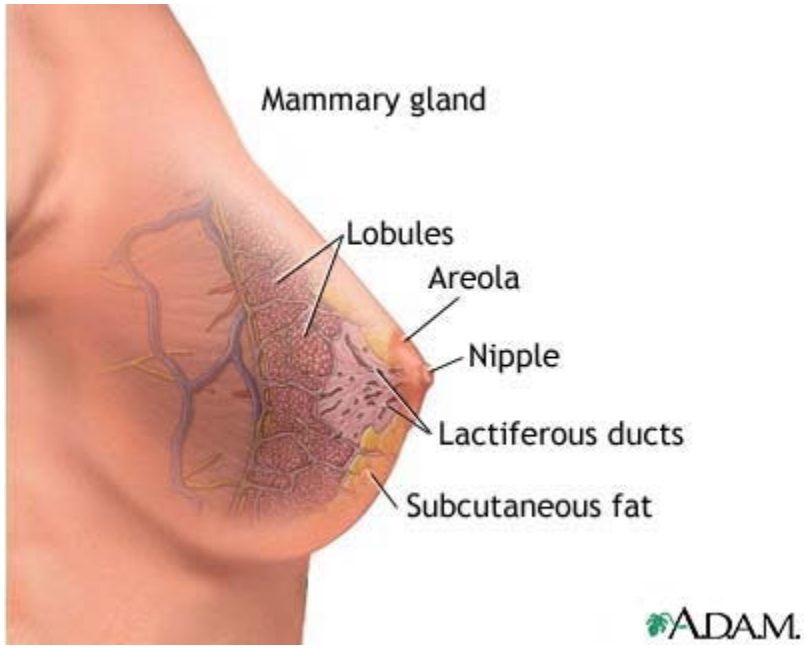





**Paget Disease of the nipple. Shows growth pattern of Paget on the pigmented portion of nipple and inside the milk duct opening**

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

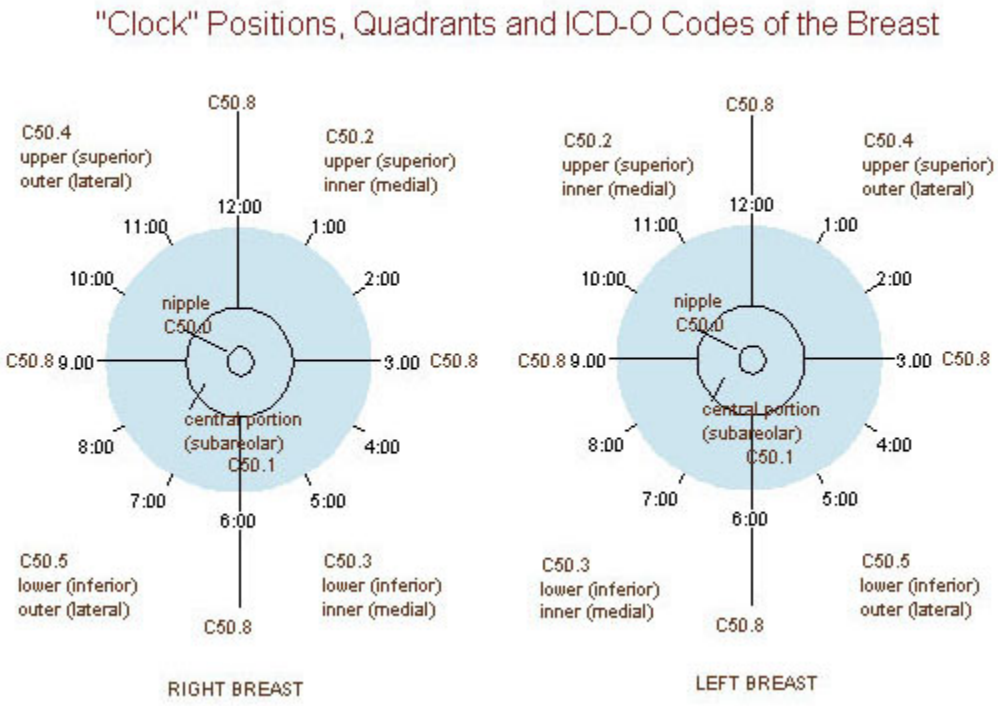
**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**



**Breast Equivalent Terms and Definitions**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 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 – M9992 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 – M9992 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 breast tumor.**

***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.

**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 – M9992 and Kaposi sarcoma M9140)**

**Rule M10** Abstract a **single primary**[i] when multiple tumors of the **same behavior** are **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 and have the ***same behavior***.

***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**

**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**

## Breast Multiple Primary Rules
## C500-C506, C508-C509
## (Excludes lymphoma and leukemia M9590 – M9992 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 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

**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.

## Breast Multiple Primary Rules
## C500-C506, C508-C509
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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.

**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.**

**Breast Histology Rules**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 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** **Table 3** or the ICD-O and all updates.

## 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 – M9992 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. 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.

## 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.

**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 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**

**Breast Histology Rules**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## Single Tumor: Invasive and In Situ Components

**Rule H8** Code the **invasive** histology when both invasive and in situ components are present.
***Note 1:*** **Ignore** the in situ term.
***Note 2:*** This is consistent with the 2007 MPH Rules.

**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**

## Single Tumor: Invasive Only

**Rule H9** 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 H10** 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).

[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 H11** 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.**

**Rule H12** 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 H13** 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 H14** 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 H15** Code duct carcinoma and invasive lobular carcinoma **8522/3** when there is both invasive carcinoma NST/duct carcinoma and invasive 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 invasive 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 H16** 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 spindle cell type 8032/3 and the remainder is metaplastic carcinoma NOS 8575/3. Code the subtype/variant: metaplastic carcinoma spindle cell type 8032/3.

**Rule H17** 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 H18** 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 H19** 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**

**Breast Histology Rules**
**C500-C506, C508-C509**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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 H20** 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 H21** 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 H22** Code Paget disease and DCIS **8543/2** when there is Paget disease (specified as **in situ**) with underlying **DCIS**.

**Rule H23** 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 H24** Code the **invasive** histology when there are invasive and in situ histologies:

- Mixed in each of multiple tumors **OR**
- In separate tumors (one or more invasive and one or more in situ)

***Example 1:*** Multiple tumors, each with invasive carcinoma NST and in situ lobular carcinoma (LCIS) mixed. Code to invasive carcinoma NST 8500/3.

***Example 2:*** One tumor is invasive carcinoma NST and the other is lobular carcinoma in situ (LCIS). Code to invasive carcinoma NST 8500/3.

**Rule H25** Code **8522** when carcinoma NST and lobular are present in multiple tumors.

- DCIS and in situ lobular **8522/2**
- Carcinoma NST/duct carcinoma and invasive lobular **8522/3**

***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 invasive lobular carcinoma 8522/3.

***Note 2:*** One tumor may be carcinoma NST and the other lobular, or all tumors may be a mixture of carcinoma NST and lobular.

***Note 3:*** This combination code specifically identifies carcinoma NST and lobular carcinoma. For all other histological combinations, continue through the rules.

***Note 4:*** These rules are hierarchical. Both histologies must be in situ or both histologies must be invasive. For example, do not use this rule for invasive carcinoma NST and in situ lobular.

**Rule H26** 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.

**Rule H27** 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 – M9992 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 – M9992 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**

## Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
## C180-C189, C199, C209
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

### 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**<br><br>***Note 1*: See Histology Rules** for instructions on coding adenocarcinoma **subtypes/variants** arising in a polyp<br><br>***Note 2*:** When the term **intestinal adenocarcinoma** is used to describe a colon primary, it simply means the **appearance** is | Adenocarcinoma, NOS<br>Adenocarcinoma/carcinoma in a polyp NOS (now coded to 8140)<br>Adenocarcinoma/carcinoma in adenomatous polyp (now coded to 8140)<br>Adenocarcinoma/carcinoma in polypoid adenoma (now coded to 8140)<br>Adenocarcinoma/carcinoma in serrated adenoma (now coded to 8140)<br>Adenocarcinoma and mucinous carcinoma, mucinous documented as less than 50% of tumor OR percentage of mucinous | Adenoid cystic carcinoma **8200**<br>Cribriform comedo-type carcinoma/ adenocarcinoma, cribriform comedo-type **8201***<br>Diffuse adenocarcinoma/carcinoma **8145**<br>High grade appendiceal mucinous neoplasm (HAMN)/Low grade appendiceal mucinous neoplasm **8480- see Note 3**<br>Linitis plastica **8142/3**Medullary adenocarcinoma/carcinoma **8510**<br>Micropapillary carcinoma **8265***<br>Mucinous/colloid adenocarcinoma/carcinoma |

| Specific and NOS Term and Code | Synonyms for Specific or NOS Term | Subtypes/Variants |
|---|---|---|
| similar to adenocarcinoma seen in the stomach and is coded to adenocarcinoma NOS **8140**<br><br>**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. | unknown/not documented<br>Adenocarcinoma and signet ring cell carcinoma, percentage of signet ring cell carcinoma documented as less than 50% of tumor OR unknown/not documented<br>Adenocarcinoma/carcinoma in tubular polyp (now coded to 8140)<br>Adenocarcinoma/carcinoma in tubulovillous polyp (now coded to 8140)<br>Adenocarcinoma/carcinoma in villous adenoma (now coded to 8140)<br>Adenocarcinoma in any type of polyp<br>Adenocarcinoma, intestinal type<br>Adenocarcinoma and cribriform carcinoma percentage of cribriform documented as less than 50% of tumor OR unknown/not documented<br>Adenocarcinoma with mucinous and signet ring cell features<br>Comedocarcinoma<br>Intestinal adenocarcinoma | **8480**<br>Mucoepidermoid carcinoma **8430**<br>Serrated adenocarcinoma **8213***<br>Signet ring cell/poorly cohesive adenocarcinoma/carcinoma **8490**<br>Superficial spreading adenocarcinoma **8143**<br>Tubulopapillary carcinoma **8263**<br>Undifferentiated adenocarcinoma/carcinoma **8020** |
| **Adenosquamous carcinoma 8560**<br>***Note:*** This code **cannot be used** for **adenocarcinoma subtypes/variants** with squamous cell/epidermoid carcinoma | Mixed adenocarcinoma NOS and epidermoid carcinoma<br>Mixed adenocarcinoma NOS and squamous cell carcinoma | |
| **Combined small cell carcinoma 8045** | Small cell carcinoma mixed with<br>• Adenocarcinoma **OR**<br>• Neuroendocrine carcinoma **OR**<br>• Any other type of carcinoma/adenocarcinoma | |
| **Gastrinoma 8153** | | |

**Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions**
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| Specific and NOS Term and Code | Synonyms for Specific or NOS Term | Subtypes/Variants |
|---|---|---|
| **Gastrointestinal stromal tumor 8936/3**<br>***Note*:** See standard setter reportability guidelines. | Gastrointestinal stromal tumor<br>GIST, NOS<br>GIST, malignant<br>Gastrointestinal stromal sarcoma | |
| **Mixed adenoneuroendocrine carcinoma 8244** | Adenocarcinoma ex-goblet cell<br>Adenocarcinoma mixed with high-grade large cell neuroendocrine carcinoma<br>Adenocarcinoma mixed with high-grade small cell neuroendocrine carcinoma<br>MANEC | Goblet cell adenocarcinoma/Goblet cell carcinoid **8243** |
| **Neuroendocrine carcinoma 8246** | NEC | Large cell NEC **8013**<br>Small cell NEC **8041** |
| **Neuroendocrine tumor Grade 1 (G1) 8240**<br>***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<br>Low-grade neuroendocrine tumor<br>NET Grade 1 (G1)<br>Well differentiated neuroendocrine tumor | EC cell serotonin-producing NET/enterochromaffin cell carcinoid **8241**<br>Neuroendocrine tumor (NET) Grade 2 (G2) **8249**<br>Somatostatin-producing NET **8156** |
| **Sarcoma NOS 8800/3** | | Angiosarcoma/hemangiosarcoma **9120/3**<br>Leiomyosarcoma **8890/3** |
| **Spindle cell carcinoma 8032** | | |
| **Squamous cell carcinoma 8070** | Epidermoid carcinoma NOS<br>Squamous cell carcinoma NOS<br>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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## 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**<br><br>***Note*:** No malignancy in polyps | Adenoma NOS | Tubular adenoma **8211/0**<br>Tubulovillous adenoma **8263/0**<br>Villous adenoma **8261/0** | Non-malignant |
| **Adenomatous polyp, high grade dysplasia 8210/2** | | | Non-reportable terminology |
| **Cowden-associated polyp No code**<br><br>***Note*:** No malignancy in polyps | Cowden disease<br>Cowden syndrome<br>Multiple hamartoma syndrome | | Non-malignant /no code |
| **Dysplasia, high grade 8148/2**<br>***Note:*** Colorectal primaries only (C180-C189, C199 and C209) | High-grade dysplasia<br>Intraepithelial neoplasia, high grade | | **CURRENTLY NOT REPORTABLE** |
| **Dysplasia, low grade 8148/0***<br>***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<br>Bussey-Garder polyposis<br>Familial multiple polyposis<br>Familiar polyposis coli<br>Familial polyposis of the colon and rectum<br>Familial polyposis of the gastrointestinal tract<br>Gardner syndrome<br>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<br>GIST, behavior not specified<br><br>***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<br>Familial juvenile polyposis<br>Generalized juvenile polyposis<br>Hamartomatous gastrointestinal polyposis;<br>Juvenile polyposis<br>Juvenile polyposis coli<br>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**<br><br>***Note*:** May have low-grade, non-invasive pseudomyxoma peritonei, mucinous implants in peritoneum or beyond | LAMN | | Non-malignant<br><br>**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 |

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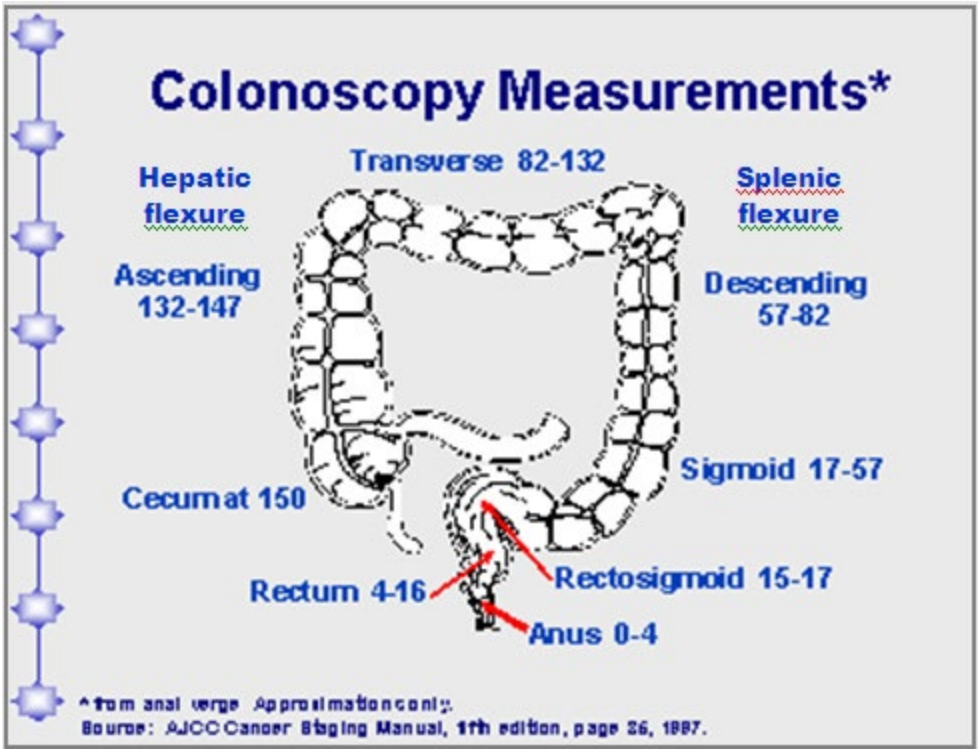
| Specific or NOS Term and Code | Synonyms | Subtype/Variant of NOS with Histology Code | Reason not reportable |
|---|---|---|---|
| **Mesenchymal tumors** | | Granular cell tumor **9580/0**<br>Hemangioma **9120/0** | Non-malignant |
| **Peutz-Jeghers polyp No code** | Intraepithelial neoplasia in Peutz-Jeghers polyp(s)<br>Periorificial lentiginosis<br>Peutz-Jeghers polyposis<br>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*** <br>***Note:*** No malignancy in polyps | Serrated polyposis<br>Sporadic serrated polyps<br>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**

## Illustrations

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



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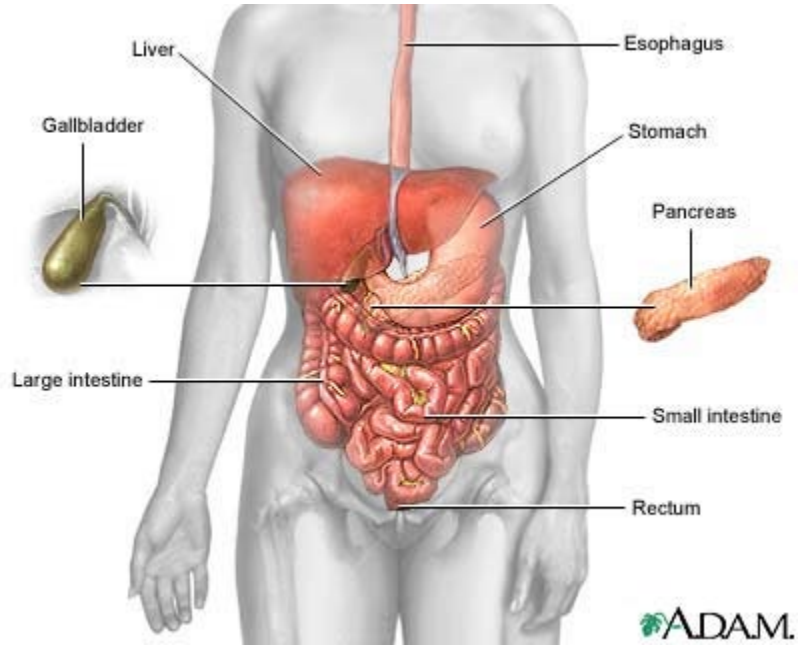
**GI System**



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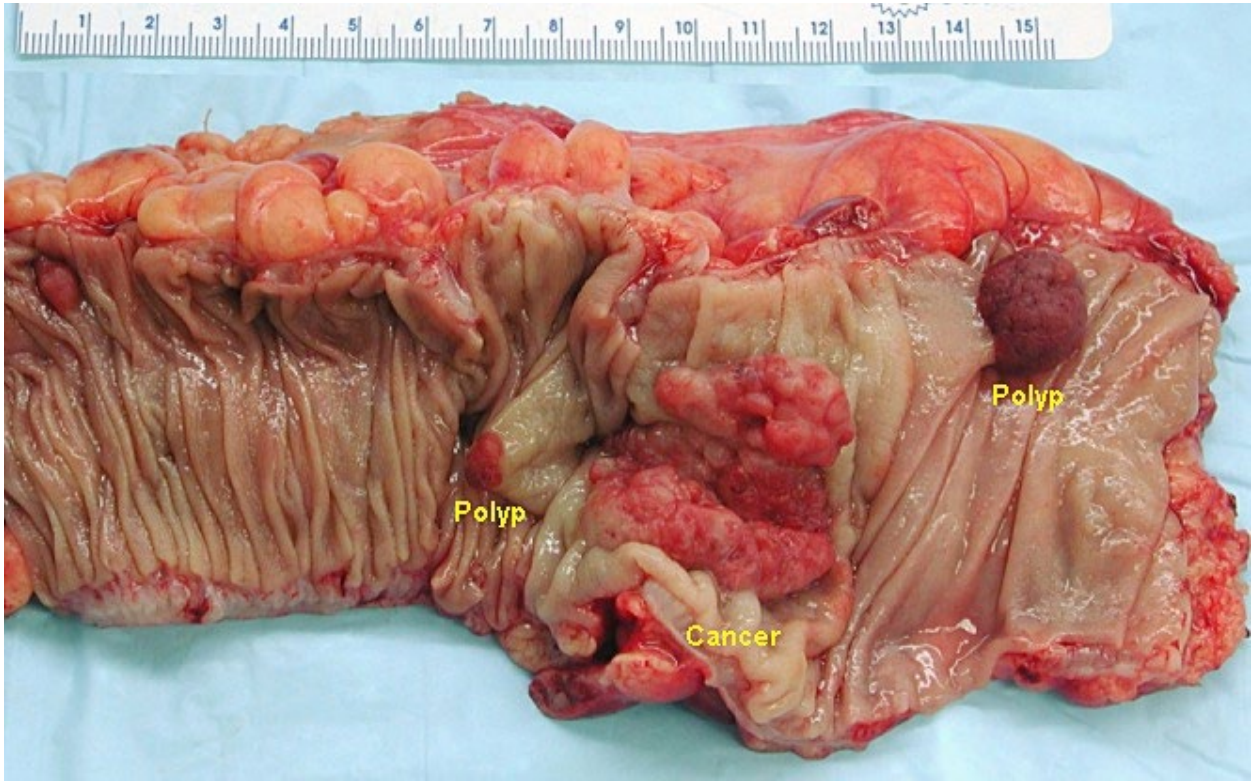
**Polyps and de novo or "frank" adenocarcinoma in colon**




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

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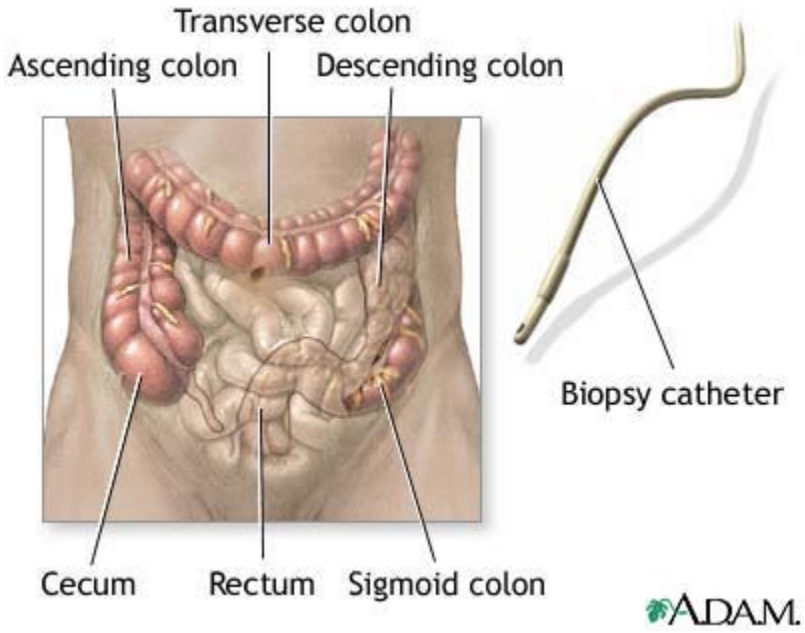
**Large intestine; snare instrument to remove polyps**



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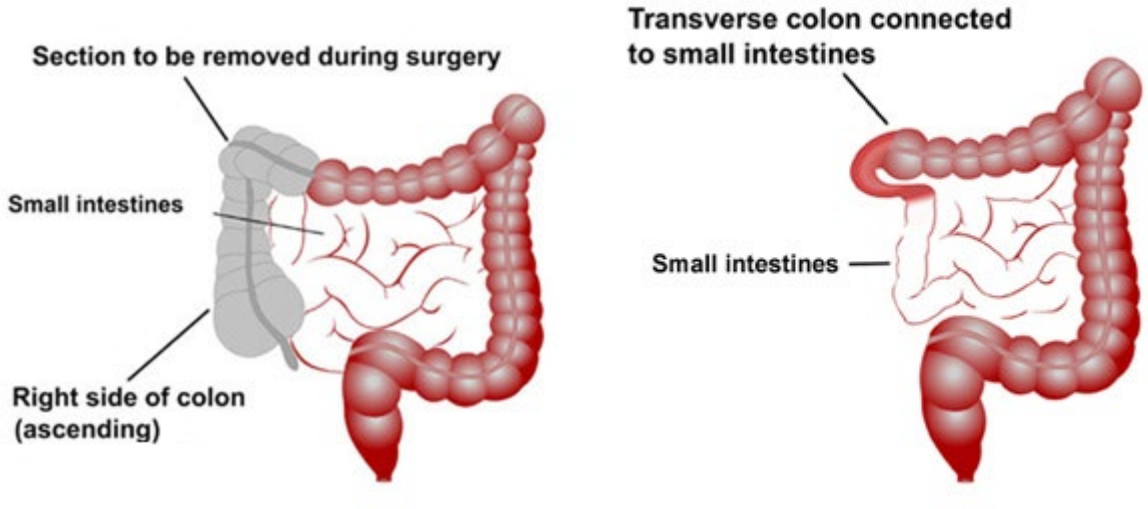
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

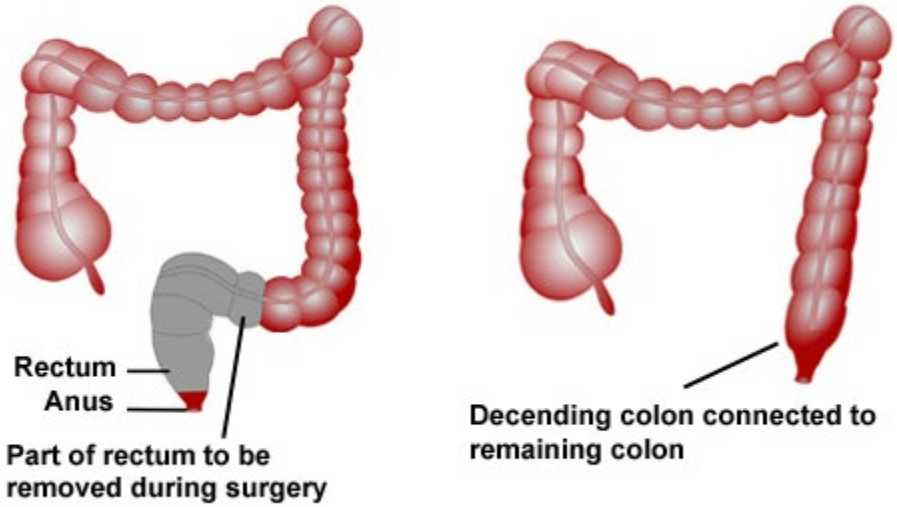
**Colon Surgery: Hemicolectomy**

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).

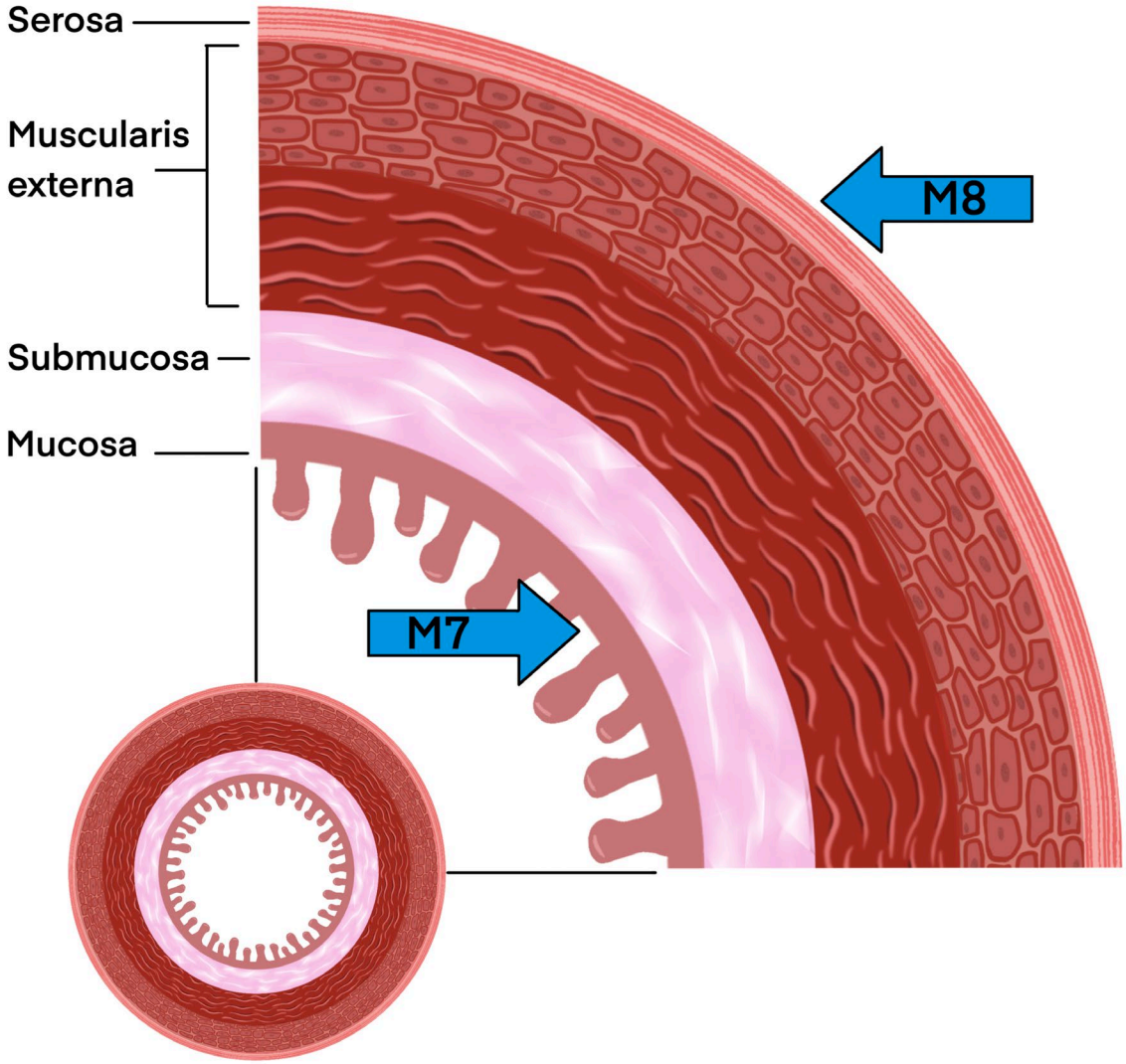



http://www.cedars-sinai.edu/Patients/Programs-and-Services/Colorectal-Cancer-Center/Services-and-Treatments/Rectal-Cancer.aspx

## Rectal Surgery



**Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions**
**C180-C189, C199, C209**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**



## Colon, Rectosigmoid, and Rectum Multiple Primary Rules
## C180-C189, C199, C209
## (Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 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 – M9992 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 – M9992 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 C187 and descending C186). **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 – M9992 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 – M9992 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 – M9992 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 – M9992 and Kaposi sarcoma M9140)**

## Coding Histology

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

1. **Code** the **most specific** histology or **subtype/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**
**C180-C189, C199, C209**
**(Excludes lymphoma and leukemia M9590 – M9992 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

***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.**

## 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
## (Excludes lymphoma and leukemia M9590 – M9992 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.

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 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.

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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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## 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

**Head and Neck Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## Table Index

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 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.

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Adenocarcinoma 8140**<br><br>***Note:*** Adenocarcinoma intestinal-type of the sinonasal tract is morphologically similar to adenocarcinomas of the intestines | Adenocarcinoma non-intestinal type<br>Low-grade adenocarcinoma<br>Renal cell-like carcinoma<br>Seromucinous adenocarcinoma<br>TAC<br>Terminal tubulous adenocarcinoma<br>Tubulopapillary low-grade adenocarcinoma | Adenocarcinoma<br>intestinal type (ITAC) **8144**<br>Colloid-type adenocarcinoma **8144**<br>Colonic-type adenocarcinoma **8144**<br>Enteric-type adenocarcinoma **8144** |
| **Lymphoepithelial carcinoma 8082** | LEC<br>Lymphoepithelioma-like carcinoma | |
| **Malignant peripheral nerve sheath tumor 9540/3** | Malignant neurilemmoma<br>Malignant schwannoma<br>MPNST<br>Neurofibrosarcoma | |
| **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<br>NUT midline carcinoma | |
| **Olfactory neuroblastoma 9522/3** | Esthesioneuroblastoma<br>Esthesioneurocytoma<br>Esthesioneuroepithelioma<br>Olfactory placode tumor<br>ONB | |

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Primitive neuroectodermal tumor 9364** | Adult neuroblastoma<br>Ewings sarcoma<br>Peripheral neuroblastoma<br>Peripheral neuroectodermal tumor<br>Peripheral neuroepithelioma | |
| **Sarcoma 8800/3**<br><br>***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.<br>***Note 2:*** Rhabdomyosarcoma **8900/3** has subtypes/variants:<br>Alveolar rhabdomyosarcoma **8920/3**<br>Embryonal rhabdomyosarcoma **8910/3**<br>Pleomorphic rhabdomyosarcoma, adult type **8901/3** | | Angiosarcoma/hemangiosarcoma **9120/3**<br>Biphenotypic sinonasal sarcoma (BSNS)/low-grade sinonasal sarcoma with neural and myogenic features **9045/3***<br>Epithelioid hemangioendothelioma **9133/3**<br>Fibrosarcoma/adult-type fibrosarcoma **8810/3**<br>Leiomyosarcoma **8890/3**<br>Malignant hemangioendothelioma **9130/3**<br>Rhabdomyosarcoma **8900/3**<br>Alveolar rhabdomyosarcoma **8920/3**<br>Embryonal rhabdomyosarcoma **8910/3**<br>Pleomorphic rhabdomyosarcoma, adult type **8901/3**<br>Spindle cell rhabdomyosarcoma **8912/3**<br>Synovial sarcoma/synovial cell sarcoma **9040/3**<br>Undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma **8802/3** |
| **Sinonasal undifferentiated carcinoma 8020**<br><br>***Note:*** This is an undifferentiated carcinoma of the Sinonasal tract. | Sinonasal carcinoma, undifferentiated<br>SNUC | |

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Squamous cell carcinoma 8070**<br><br>***Note:*** Sinonasal squamous cell tumors account for about 3% of head and neck malignancies. | Squamous cell carcinoma, usual type 8070/3<br>Conventional Squamous cell carcinoma NOS<br>Epidermoid carcinoma, NOS 8070/3<br>Epidermoid carcinoma in situ, NOS 8070/2<br>Squamous carcinoma 8070/3<br>Squamous cell carcinoma in situ, NOS 8070/2<br>Squamous cell epithelioma 8070/3<br>Intraepithelial squamous cell carcinoma 8070/2 | Keratinizing squamous cell carcinoma (KSCC) **8071**<br>Epidermoid carcinoma, keratinizing<br>Squamous cell carcinoma, large cell, keratinizing<br>Squamous cell carcinoma, large cell, nonkeratinizing/Squamous cell carcinoma, nonkeratinizing, NOS **8072**<br>Schneiderian carcinoma/cylindrical cell carcinoma **8121**<br>Sarcomatoid squamous cell carcinoma/spindle cell squamous cell carcinoma (SC-SCC) **8074** |
| **Teratocarcinosarcoma 9081** | Blastoma<br>Malignant teratoma<br>Teratocarcinoma<br>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**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 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**<br>Keratinizing squamous cell carcinoma **8071**<br>Non-keratinizing squamous cell carcinoma **8072**<br>Lymphoepithelial carcinoma **8082**<br>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.

**Head and Neck Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 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** | Chondrosarcoma grade 2/3<br>Chondrosarcoma NOS | |
| **Liposarcoma 8850** | Atypical lipomatous tumor<br>Well-differentiated liposarcoma | |
| **Squamous cell carcinoma (SCC) 8070** | Epidermoid carcinoma<br>Conventional Squamous cell carcinoma NOS | Adenosquamous carcinoma (ASC) **8560**<br>Basaloid squamous cell carcinoma (BSCC) **8083**<br>Lymphoepithelial carcinoma (LEC)/lymphoepithelioma-like carcinoma **8082**<br>Keratinizing squamous cell carcinoma **8071**<br>Non-keratinizing squamous cell carcinoma **8072**<br>Papillary squamous cell carcinoma (PSCC) **8052**<br>Spindle cell squamous cell carcinoma (SC-SCC) **8074**<br>Verrucous squamous cell carcinoma (VC) **8051** |
| **Well-differentiated neuroendocrine carcinoma 8240** | Carcinoid<br>Neuroendocrine carcinoma grade 1 | Large cell neuroendocrine carcinoma/LCNEC **8013**<br>Neuroendocrine carcinoma grade 2/moderately-differentiated neuroendocrine carcinoma/atypical carcinoid **8249**<br>Small cell neuroendocrine carcinoma/small cell carcinoma/SmCC **8041** |

## 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<br>Squamous carcinoma<br>Squamous cell carcinoma NOS | Acantholytic squamous cell carcinoma **8075**<br>Keratinizing squamous cell carcinoma **8071**<br>Non-keratinizing squamous cell carcinoma **8072** |

*** 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).

**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**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Adenoid cystic carcinoma 8200** | | |
| **Polymorphous adenocarcinoma 8525** | Cribriform adenocarcinoma<br>Polymorphous low-grade adenocarcinoma<br>Terminal duct carcinoma | |
| **Squamous cell carcinoma 8070**<br><br>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.<br><br>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 | Keratinizing squamous cell carcinoma **8071 (see note 1)**<br>Non-keratinizing squamous cell carcinoma **8072 (see note 2)**<br>Squamous cell carcinoma HPV-negative **8086***<br>**Cases diagnosed prior to 1/1/2022:**<br>***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.<br>**Cases diagnosed 1/1/2022 forward:**<br>***Note:*** HPV mediated (p16-) test results can be used to assign code 8086.<br><br>Squamous cell carcinoma HPV-positive **8085***<br>**Cases diagnosed prior to 1/1/2022:** |

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| | | ***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.<br>**Cases diagnosed 1/1/2022 forward:**<br>***Note:*** HPV mediated (p16+) test results can be used to assign code 8085. |

*** 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**
**(Excludes lymphoma and leukemia M9590 – M9992 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**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

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

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Adenoid cystic carcinoma 8200** | ACC | |
| **Carcinosarcoma 8980** | Carcinosarcoma NOS<br>True malignant mixed tumor | |
| **Cystadenocarcinoma 8440** | | |
| **Epithelial-myoepithelial carcinoma 8562** | Adenomyoepithelioma | |
| **Lymphoepithelial carcinoma (LEC) 8082** | Lymphoepithelioma-like carcinoma<br>Malignant lymphoepithelial lesion<br>Undifferentiated carcinoma with lymphoid stroma | |
| **Myoepithelial carcinoma 8982** | Malignant myoepithelioma | |
| **Neuroendocrine carcinoma 8246** | Neuroendocrine carcinoma NOS | Large-cell neuroendocrine carcinoma **8013**<br>Small cell carcinoma NOS/small cell neuroendocrine carcinoma **8041** |
| **Oncocytic carcinoma 8290** | Malignant oncocytoma<br>Oncocytic adenocarcinoma | |
| **Sebaceous adenocarcinoma 8410** | Sebaceous carcinoma. NOS | |
| **Secretory carcinoma 8502*** | Mammary analog secretory carcinoma | |
| **Squamous cell carcinoma 8070** | Conventional Squamous cell carcinoma NOS<br>SCC<br>Squamous carcinoma<br>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<br>Ameloblastic carcinoma<br>Ameloblastic carcinoma, dedifferentiated<br>Ameloblastic carcinoma, secondary type<br>Primary intraosseous carcinoma NOS (PIOC)<br>Sclerosing odontogenic carcinoma (SOC) | Metastasizing ameloblastoma **9310/3**<br><br>***Note:*** This is an ameloblastoma which has a benign appearance but metastasizes |
| **Clear cell odontogenic carcinoma 9341***<br><br>***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<br>Calcifying ghost cell odontogenic carcinoma<br>Carcinoma arising in calcifying odontogenic cyst<br>Malignant calcifying ghost cell odontogenic tumor<br>Malignant calcifying odontogenic cyst<br>Malignant epithelial odontogenic ghost cell tumor | |

| Specific or NOS Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| **Odontogenic carcinosarcoma 8980/3** | Ameloblastic carcinosarcoma<br>Malignant odontogenic mixed tumor<br>Mixed odontogenic carcinoma | Odontogenic sarcoma/ameloblastic fibrosarcoma **9330/3** |
| **Sarcoma NOS 8800/3**<br><br>*Note 1:* Osteosarcoma **9180/3** has subtypes/variants:<br>Chondroblastic osteosarcoma **9181/3**<br>Intraosseous well-differentiated osteosarcoma/low-grade central osteosarcoma **9187/3**<br>Parosteal osteosarcoma **9192/3**<br>Periosteal osteosarcoma **9193/3**<br><br>*Note 2:* Chondrosarcoma grade 2/3 **9920/3** has a subtype/ variant:<br>Mesenchymal chondrosarcoma **9240/3** | | Chondrosarcoma grade 2/3 **9220/3**<br>Mesenchymal chondrosarcoma **9240/3**<br>Osteosarcoma/osteogenic sarcoma **9180/3**<br>Chondroblastic osteosarcoma **9181/3**<br>Intraosseous well-differentiated osteosarcoma/low-grade central osteosarcoma **9187/3**<br>Parosteal osteosarcoma **9192/3**<br>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**<br><br>***Note:*** The endolymphatic sac is located within the inner ear C301. | Adenocarcinoma<br>Heftner tumor<br>Low-grade papillary adenocarcinoma of endolymphatic sac origin |
| **Squamous cell carcinoma of the middle ear 8070**<br><br>***Note:*** This neoplasm **arises** in the squamous epithelium within the middle ear C301. | SCC<br>Squamous carcinoma<br>Squamous cell carcinoma NOS |

## Table 9: Paraganglioma of Carotid Body, 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.

Cases diagnosed prior to 1/1/2021:
Only report these neoplasms when the pathology/tissue specifies malignant behavior /3. 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.

The primary site for paragangliomas is the 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 and code for specific histologies which do not have subtypes/variants.
**Column 2** lists **synonyms** for the specific term. Synonyms have the **same ICD-O code** as the specific term.

**Table begins on next page**

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific Term and Code | Synonyms for Specific Histology |
|---|---|
| **Carotid body paraganglioma 8690/3**<br>**Cases diagnosed prior to 1/1/2021:**<br>***Note 1:*** This neoplasm is only reportable when documented as malignant/invasive /3 behavior.<br>***Note 2:*** Cases diagnosed as malignant prior to 1/1/2021 should be reported as 8692/3.<br>**Cases diagnosed 1/1/2021 forward:**<br>***Note 1***: The term "malignant" is no longer required to assign /3.<br>***Note 2***: Cases diagnosed 1/1/2021 forward are coded 8692/3 per ICD-O-3.2. | Carotid body tumor<br>Chemodectoma, carotid<br>Non-chromaffin paraganglioma, carotid |
| **Laryngeal paraganglioma 8690/3**<br>**Cases diagnosed prior to 1/1/2021:**<br>***Note 1:*** This neoplasm is only reportable when documented as malignant/invasive /3 behavior.<br>***Note 2:*** Cases diagnosed as malignant prior to 1/1/2021 should be reported as 8690/3.<br>**Cases diagnosed 1/1/2021 forward:**<br>Note 1: The term "malignant" is no longer required to assign /3.<br>Note 2: Cases diagnosed 1/1/2021 forward are coded 8693/3 per ICD-O-3.2.<br>***Note 3:*** Vagal paraganglioma has the same proposed histology code as laryngeal paraganglioma. Laryngeal and vagal are in separate rows to emphasize the primary site. | Chemodectoma, laryngeal<br>Non-chromaffin paraganglioma, laryngeal |
| **Middle ear paraganglioma 8690/3**<br>**Cases diagnosed prior to 1/1/2021:**<br>***Note 1:*** This neoplasm is only reportable when documented as malignant/invasive /3 behavior.<br>***Note 2:*** Cases diagnosed as malignant in 2018 should be reported as 8690/3.<br>**Cases diagnosed 1/1/2021 forward:**<br>**Note:** The term "malignant" is no longer required to assign /3. | Glomus jugulare tumor of middle ear<br>Glomus tympanicum<br>Jugulotympanic chemodectoma |
| **Paraganglioma, NOS 8680/3**<br>**Cases diagnosed prior to 1/1/2021:**<br>***Note*:** This neoplasm is reportable only when documented as malignant/invasive /3 behavior<br>**Cases diagnosed 1/1/2021 forward:**<br>***Note*:** The term "malignant" is n longer required to assign /3. | |

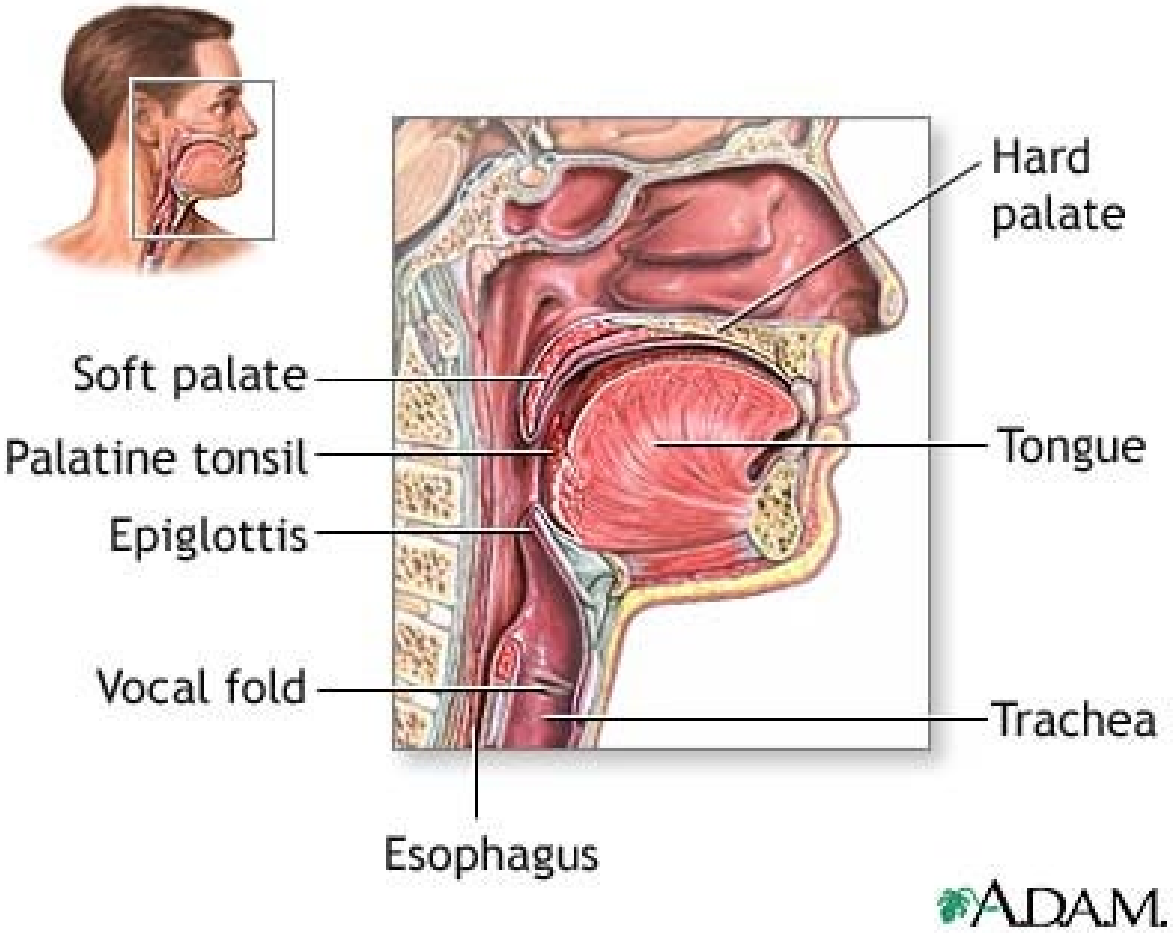
| Specific Term and Code | Synonyms for Specific Histology |
|---|---|
| **Vagal paraganglioma 8690/3**<br>**Cases diagnosed prior to 1/1/2021:**<br>***Note 1:*** This neoplasm is only reportable when documented as malignant/invasive /3 behavior.<br>***Note 2:*** Cases diagnosed as malignant prior to 1/1/2021 should be reported as 8690/3.<br>**Cases diagnosed 1/1/2021 forward:**<br>***Note 1*:** The term "malignant" is no longer required to assign /3.<br>***Note 2*:** Cases diagnosed 1/1/2021 forward are coded 8693/3 per ICD-O-3.2<br>***Note 3:*** Vagal paraganglioma has the same proposed histology code as laryngeal paraganglioma. Laryngeal and vagal are in separate rows to emphasize the primary site. | Glomus jugulare tumor of vagal trunk<br>Chemodectoma of vagal trunk<br>Non-chromaffin paraganglioma of vagal trunk |

## Table 10: Paired Sites

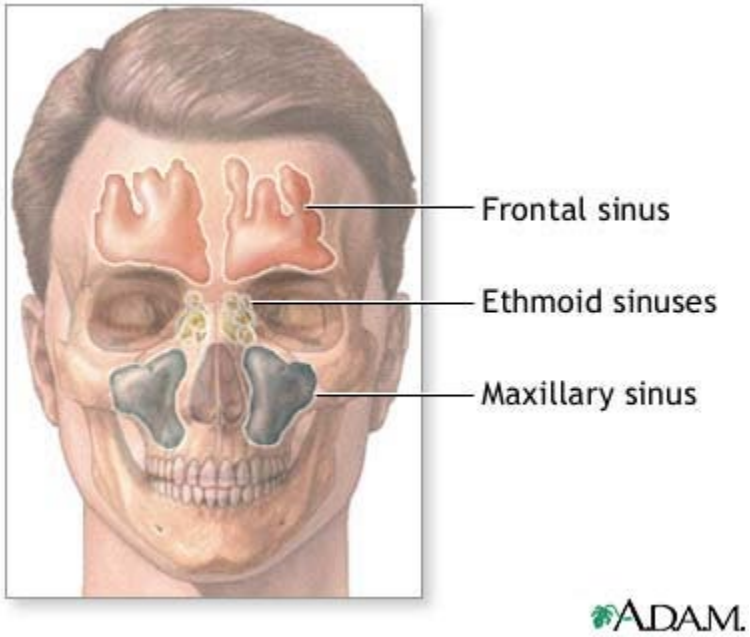
Laterality **must be coded** for all of the following sites. SEER does allow coding laterality for sites not listed in **Table 10.**

| Paired Sites | Site Code |
|---|---|
| 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 |

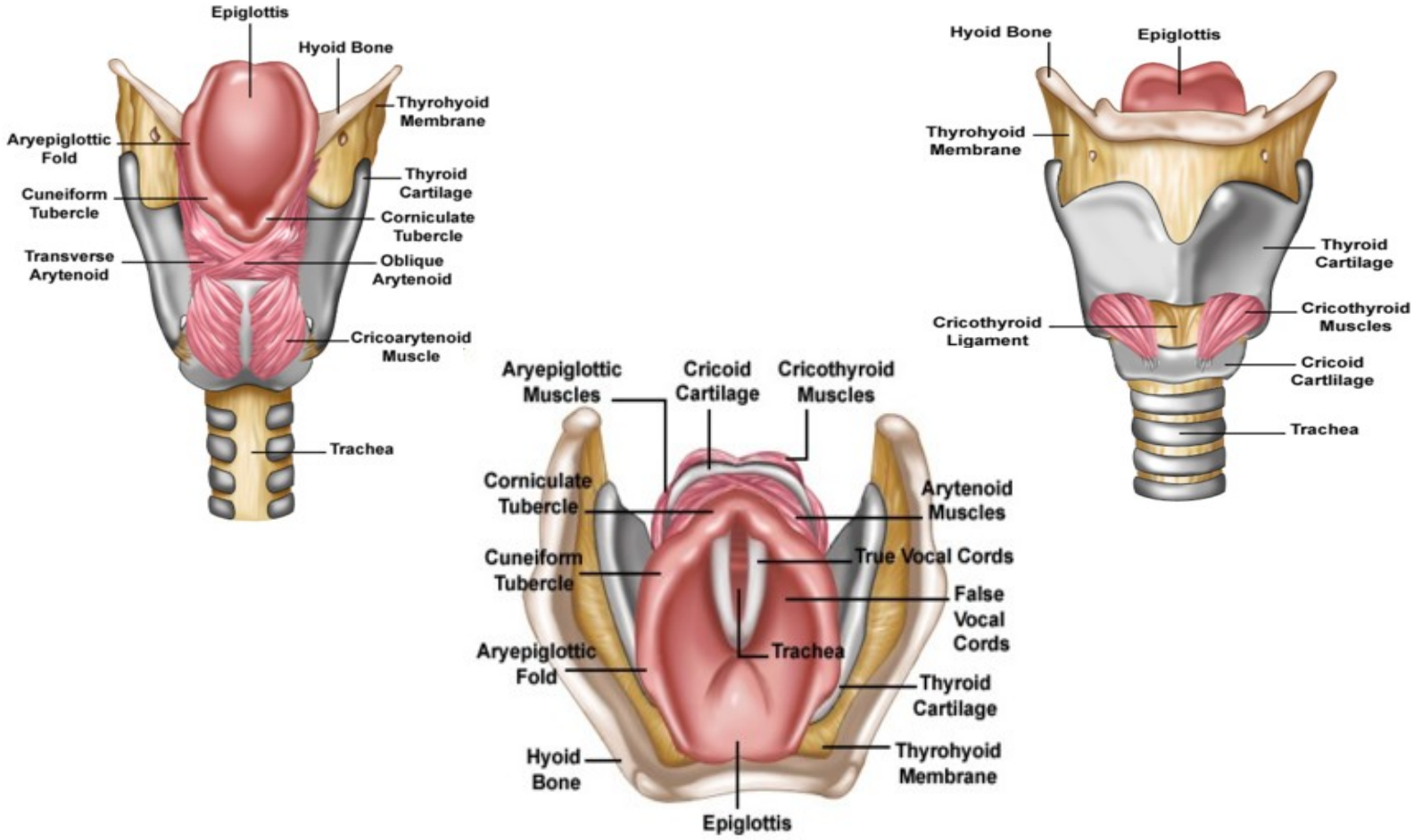
## Illustrations



**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**




Nasal Sinuses




Larynx

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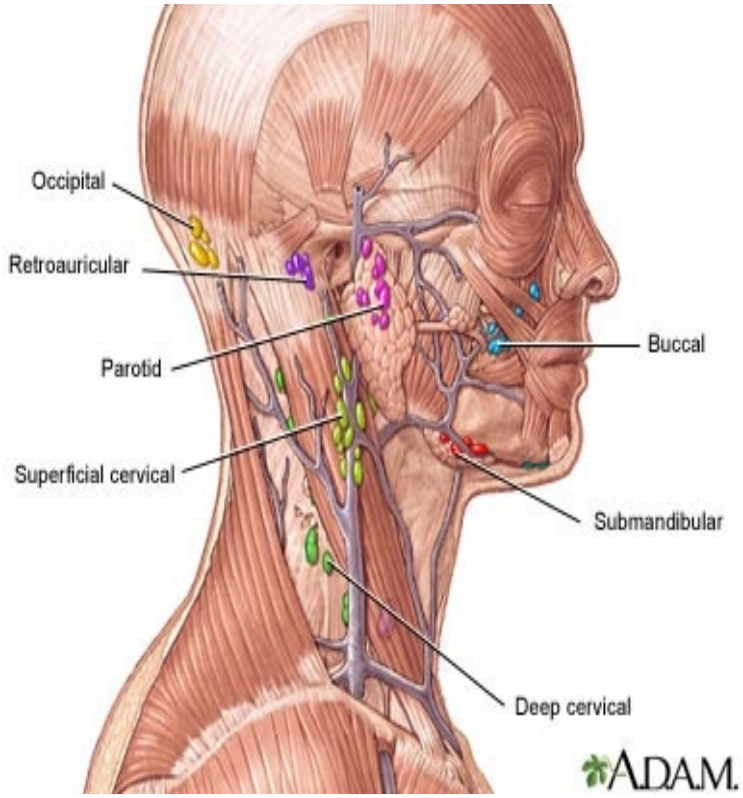
**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**



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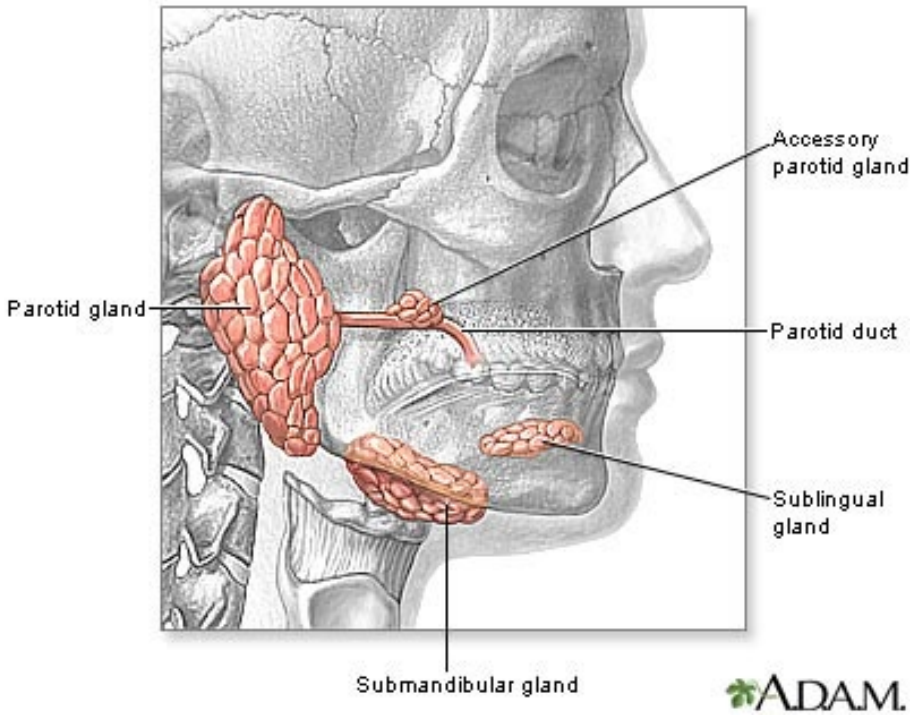

**Head and Neck Equivalent Terms and Definitions**
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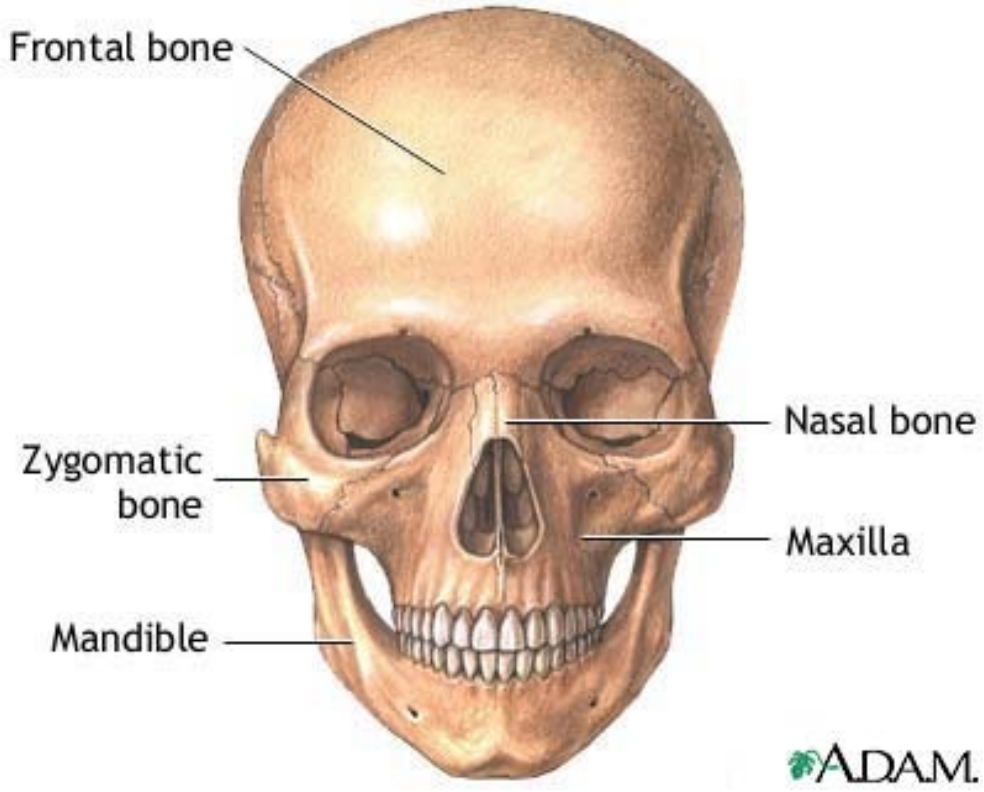

**Head and Neck Equivalent Terms and Definitions**
**C000-C148, C300-C339, C410, C411, C479**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

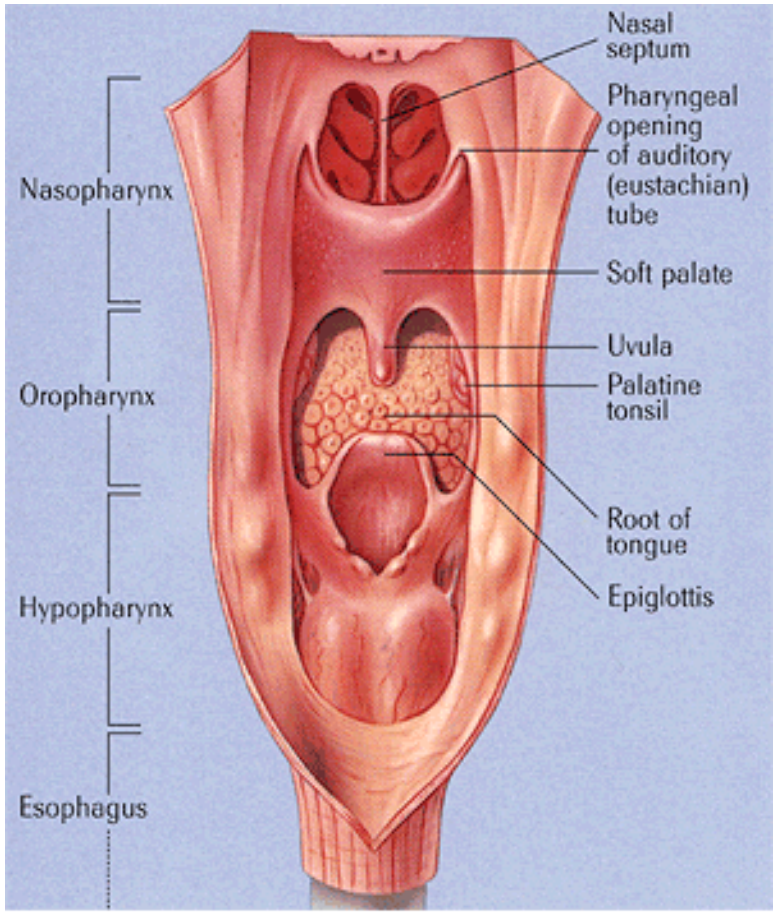


Image made available by a generous grant from Bristol-Myers Squibb

## Head and Neck Multiple Primary Rules
## C000-C148, C300-C339, C410, C411, C479
## (Excludes lymphoma and leukemia M9590 – M9992 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
## (Excludes lymphoma and leukemia M9590 – M9992 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 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 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:*** 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
## (Excludes lymphoma and leukemia M9590 – M9992 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**
**(Excludes lymphoma and leukemia M9590 – M9992 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**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## Coding Histology

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

1. **Code** the **most specific** histology or **subtype/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
## (Excludes lymphoma and leukemia M9590 – M9992 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:*** **HPV-positive is not equivalent to HPV-mediated (p16+).** 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.

**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, squamous cell carcinoma **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 – M9992 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.

## 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.

## 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
- 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

**Kidney Equivalent Terms and Definitions**
**C649**
**(Excludes lymphoma and leukemia M9590 – M9992 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**
- **Phenotype** is not equivalent to **subtype/type/variant**

## 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.

**Kidney Equivalent Terms and Definitions**
**C649**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| NOS/Specific Histology Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| Nephroblastoma **8960** | Wilms tumor | |
| Neuroendocrine tumor (NET) **8240** | Carcinoid [OBS]<br>Well-differentiated neuroendocrine tumor | Large cell neuroendocrine carcinoma/tumor **8013**<br>Small cell neuroendocrine carcinoma **8041** |
| Renal cell carcinoma NOS **8312**<br><br>***Note 1:*** WHO, IARC, and CAP agree that sarcomatoid carcinoma is a pattern of differentiation, not a specific subtype, of renal cell carcinoma.<br><br>***Note 2:*** Sarcomatoid is listed in the CAP Kidney protocol under the header "features."<br><br>***Note 3:*** Continue coding sarcomatoid renal cell carcinoma as 8312 until otherwise indicated. | RCC<br>Sarcomatoid carcinoma<br>Sarcomatoid renal cell carcinoma<br>Succinate dehydrogenase-deficient renal cell carcinoma (SDHD)<br>Unclassified renal cell carcinoma | Acquired cystic disease-associated renal cell carcinoma/tubulocystic renal cell carcinoma **8316***<br>Chromophobe renal cell carcinoma (ChRCC) **8317**<br>Clear cell papillary renal cell carcinoma **8323/3**<br>***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.**<br>Clear cell renal cell carcinoma (ccRCC) **8310**<br>Collecting duct carcinoma **8319**<br>Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma **8311***<br>MiT family translocation renal cell carcinomas **8311***<br>Succinate dehydrogenase-deficient renal cell carcinoma (SDHS) **8311* (reportable beginning 1/1/2022)**<br>***Note*:** Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma , MiT family translocation renal cell carcinomas, and succinate dehydrogenase-deficient renal cell carcinomas 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).<br>Mucinous tubular and spindle cell carcinoma **8480***<br>Papillary renal cell carcinoma (PRCC) **8260** |

**Kidney Equivalent Terms and Definitions**
**C649**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| NOS/Specific Histology Term and Code | Synonyms | Subtypes/Variants |
|---|---|---|
| | | Renal medullary carcinoma **8510*** <br>***Note*:** This is a **new** term (previously called renal spindle cell carcinoma). |
| Sarcoma **8800/3** <br><br>***Note*: Rhabdomyosarcoma** is a NOS with the following subtype/variants: <br>Alveolar rhabdomyosarcoma **8920** <br>Embryonal rhabdomyosarcoma **8910** <br>Pleomorphic rhabdomyosarcoma **8901** <br>Spindle cell/sclerosing rhabdomyosarcoma **8912** | | Angiosarcoma **9120/3** <br>Clear cell sarcoma/bone-metastasizing renal tumor of childhood **8964/3** <br>Leiomyosarcoma/renal vein leiomyosarcoma **8890/3** <br>Osteosarcoma **9180/3** <br>Primitive/peripheral neuroectodermal tumor (pNET)/Ewing sarcoma **9364/3** <br>Rhabdomyosarcoma **8900/3** <br>Alveolar rhabdomyosarcoma **8920/3** <br>Embryonal rhabdomyosarcoma **8910/3** <br>Pleomorphic rhabdomyosarcoma **8901/3** <br>Spindle cell/sclerosing rhabdomyosarcoma **8912/3** <br>Synovial sarcoma **9040/3** |

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

**Kidney Equivalent Terms and Definitions**
**C649**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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.

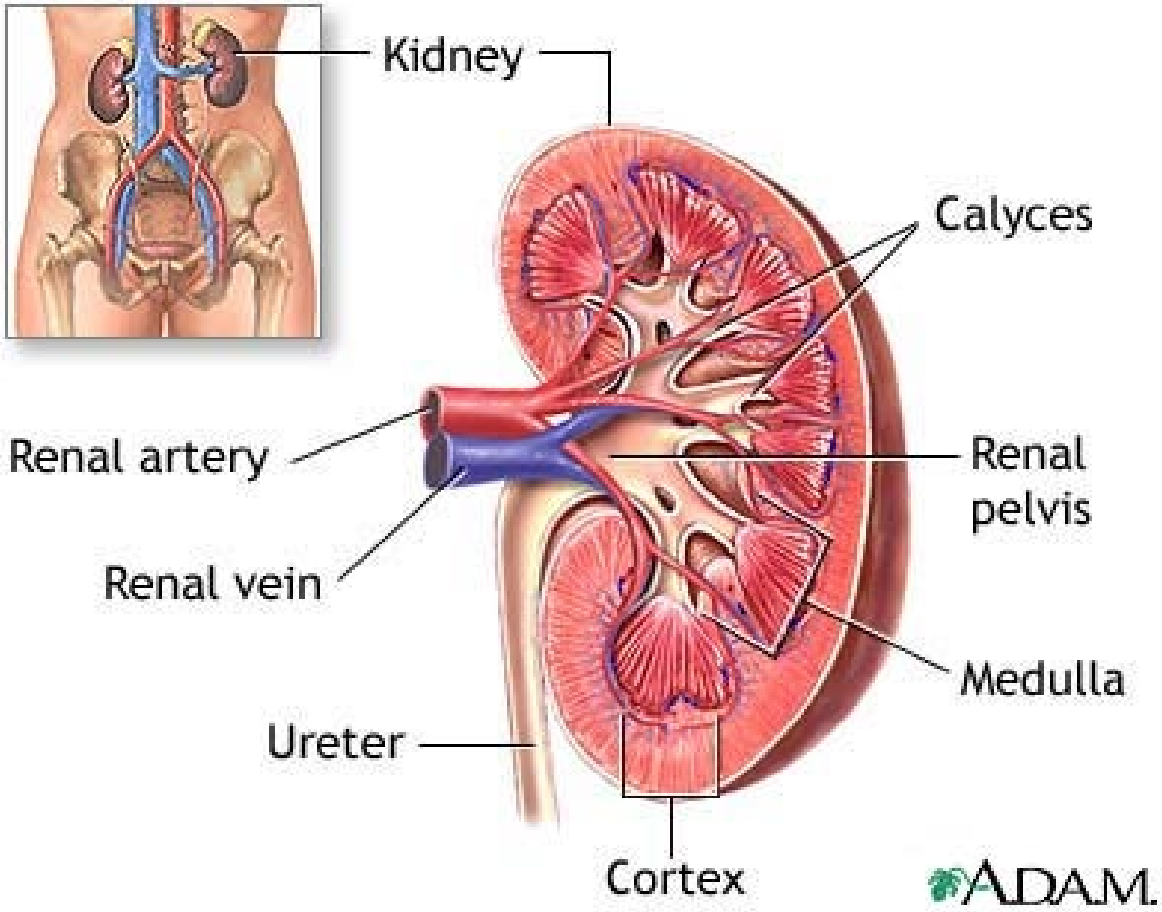
| Not Reportable Histology Term and Code | Synonyms |
|---|---|
| Adult cystic teratoma **8959/0** | Mixed epithelial and stromal tumor<br>Renal epithelial stromal tumor |
| Angiomyolipoma **8860/0** | |
| Congenital mesoblastic nephroma **8960/1** | CMN |
| Cystic partially-differentiated nephroblastoma **8959/1** | |
| Epithelioid angiolipoma **8860/1*** | |
| Hemangioblastoma **9161/1** | |
| Hemangioma **9120/0** | |
| Juxtaglomerular cell tumor **8361/0** | |
| 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)** | |
| Oncocytoma **8290/0** | |
| Papillary adenoma **8260/0** | |
| Paraganglioma **8700/0** | 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 – M9992 and Kaposi sarcoma M9140)**

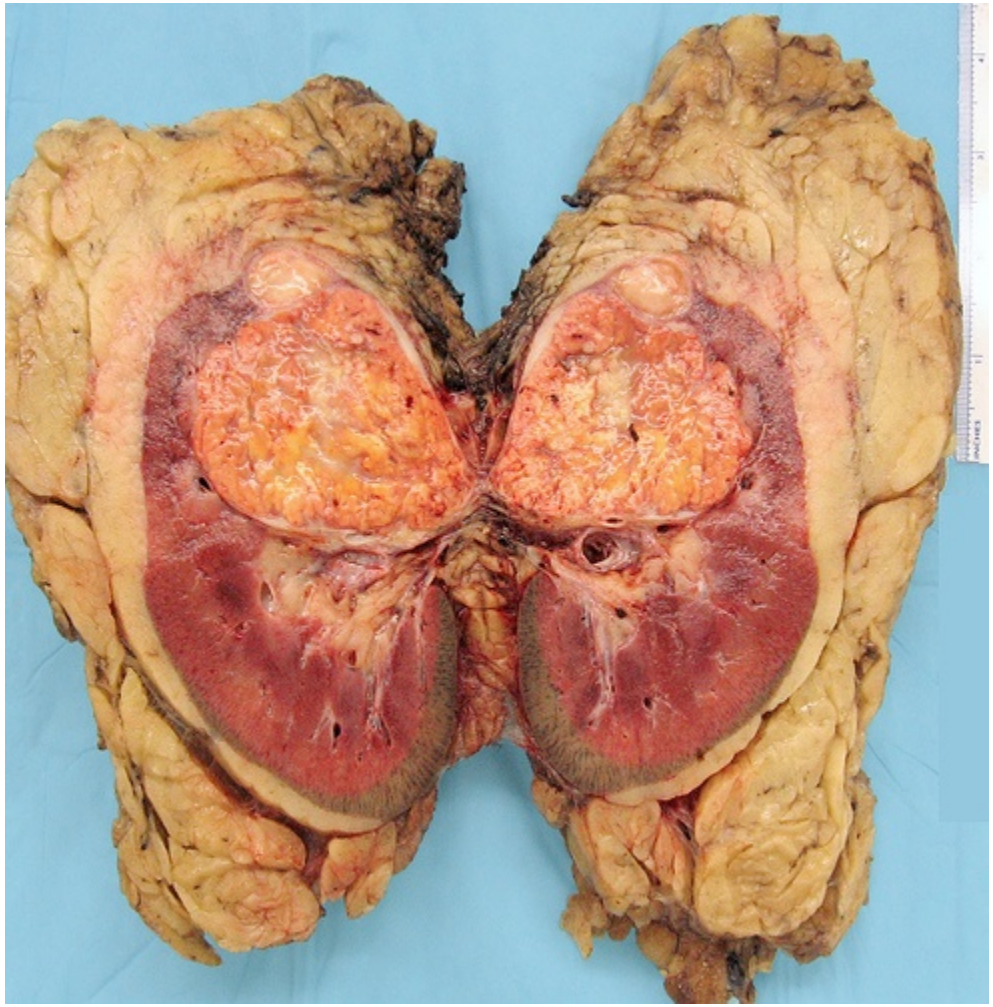
## Illustrations

**Kidney Anatomy (Includes Renal Pelvis)**



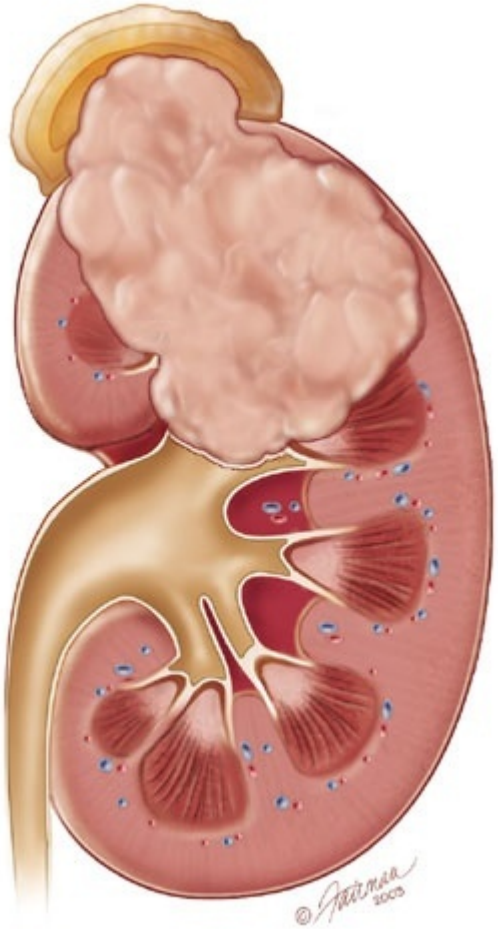
**Kidney Equivalent Terms and Definitions**
**C649**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

**Pathology Specimen Kidneys**



**Kidney Equivalent Terms and Definitions**
**C649**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## Kidney Cancer



# Kidney Multiple Primary Rules
# C649
# (Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 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).

**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.

## Kidney Multiple Primary Rules
## C649
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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 evidence** 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.

**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):

- MiT family translocation renal cell carcinoma and Hereditary leiomyomatosis
- MiT family translocation renal cell carcinoma and Renal cell carcinoma-associated renal cell carcinoma
- MiT family translocation renal cell carcinoma and Succinate dehydrogenase-deficient renal cell carcinoma (SDHS)
- Hereditary leiomyomatosis and Succinate dehydrogenase-deficient renal cell carcinoma (SDHS)
- Renal cell carcinoma-associated renal cell carcinoma and Succinate dehydrogenase-deficient renal cell carcinoma (SDHS)

## Kidney Multiple Primary Rules
## C649
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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

**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.

## Kidney Multiple Primary Rules
## C649
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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.**

**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 – M9992 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 – M9992 and Kaposi sarcoma M9140)**

## Coding Histology

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

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

   ***Example 1:*** Diagnosis for a single tumor is 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.**

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 – M9992 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 – M9992 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 – M9992 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
- 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
- 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 – M9992 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
- **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**

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Terminology | Laterality | Site Term and Code |
|---|---|---|
| **Bronchus intermedius**<br>**Carina**<br>**Hilus of lung**<br>**Perihilar** | Bilateral | Mainstem bronchus **C340**<br>*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**<br>**Apex of lung**<br>**Lung apex**<br>**Pancoast tumor**<br>**Superior lobar bronchus**<br>**Upper lobe bronchi** | Bilateral | Upper lobe **C341** |
| **Middle lobe**<br>**Middle lobe bronchi** | Right | Middle lobe **C342** |
| **Base of lung**<br>**Lower lobar bronchus**<br>**Lower lobe**<br>**Lower lobe bronchi**<br>**Lower lobe segmental bronchi** | Bilateral | Lower lobe **C343** |
| **Overlapping lesion of lung** | Bilateral | Overlapping lesion of lung **C348**<br>*Note*: **One** lesion/tumor which **overlaps two or more** lobes |

**Table continues on next page**

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Terminology | Laterality | Site Term and Code |
|---|---|---|
| **Bronchus NOS**<br>**Bronchogenic**<br>**Extending up to the hilum**<br>**Extending down to the hilar region**<br>**Lung NOS**<br>**Pulmonary NOS**<br>**Suprahilar NOS** | Bilateral | Lung NOS **C349**<br>***Note***: Includes<br>• Multiple tumors in different lobes of ipsilateral lung **OR**<br>• Multiple tumors in ipsilateral lung; unknown if same lobe or different lobe **OR**<br>• Tumor in bronchus, unknown if mainstem or lobar bronchus **OR**<br>• Tumor present, unknown which lobe |
| **Lobar bronchi NOS**<br>**Lobar bronchus NOS** | Bilateral | Code the lobe in which the lobar bronchus tumor is present **C34__**<br>***Note***: When **lobe** of origin is **not documented/unknown**, **code** to lung NOS **C349** |

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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 – M9992 and Kaposi sarcoma M9140)**

| Required Terms | Combination Histologies and Code |
|---|---|
| Adenocarcinoma NOS<br>**AND**<br>Squamous cell carcinoma NOS<br><br>***Note*:** Diagnosis **must be** adenocarcinoma NOS and squamous cell carcinoma NOS, **NOT** any of the **subtypes/variants** of adenocarcinoma or squamous cell carcinoma | Adenosquamous carcinoma **8560** |
| Giant cell carcinoma<br>**AND**<br>Spindle cell carcinoma<br><br>***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**<br><br>***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<br>**AND**<br>Myoepithelial carcinoma | Epithelial-myoepithelial carcinoma **8562** |
| Large cell neuroendocrine carcinoma<br>**AND**<br>Adenocarcinoma NOS **OR**<br>Squamous cell carcinoma NOS **OR**<br>Spindle cell carcinoma **OR**<br>Giant cell carcinoma | Combined large cell neuroendocrine carcinoma **8013** |
| Mucinous carcinoma, invasive<br>**AND**<br>Non-mucinous carcinoma, invasive | Mixed invasive mucinous and non-mucinous carcinoma **8254/3*** |

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Required Terms | Combination Histologies and Code |
|---|---|
| Small cell carcinoma/neuroendocrine tumor (NET)<br>***Note:*** **Includes subtypes/variants** of small cell/neuroendocrine tumor. See Table 3 for subtypes/variants.<br><br>**AND**<br><br>**At least one** of the following:<br>• Adenocarcinoma and any subtype/variant of adenocarcinoma<br>• Adenosquamous carcinoma<br>• Large cell carcinoma and any subtype/variant of large cell carcinoma (includes large cell neuroendocrine carcinoma)<br>• Squamous cell carcinoma and any subtype/variant of squamous cell carcinoma<br>• Non-small cell carcinoma | Combined small cell carcinoma **8045** |
| Squamous cell carcinoma (epidermoid carcinoma)<br><br>**AND**<br><br>Large cell non-keratinizing squamous cell carcinoma<br><br>***Note:*** Squamous cell carcinoma and epidermoid carcinoma are synonyms | Squamous cell carcinoma, large cell, nonkeratinizing **8072** |
| Squamous cell carcinoma (epidermoid carcinoma)<br><br>**AND**<br><br>Small cell nonkeratinizing squamous cell carcinoma<br><br>***Note:*** Squamous cell carcinoma and epidermoid carcinoma are synonyms | Squamous cell carcinoma, small cell, nonkeratinizing **8073** |

**Lung Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Required Terms | Combination Histologies and Code |
|---|---|
| Squamous cell carcinoma, keratinizing<br><br>**AND**<br><br>Squamous cell carcinoma, non-keratinizing | Squamous cell carcinoma, NOS **8070** |
| Squamous cell (epidermoid) carcinoma<br><br>**AND**<br><br>**One or both** of the following:<br>• Sarcomatoid carcinoma<br>• Spindle cell carcinoma<br><br>***Note 1:*** **Does not include subtypes/variants** of squamous cell. See Table 3 for subtypes/variants.<br>***Note 2:*** Squamous cell carcinoma and epidermoid carcinoma are synonyms. | Squamous cell carcinoma, sarcomatoid **8074**<br>Squamous cell carcinoma, spindle cell **8074** |

**Table continues on next page**

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Required Terms | Combination Histologies and Code |
|---|---|
| Diagnosis must be a single tumor which meets one of the following two criteria:<br>1. At least two of the subtypes/variants of adenocarcinoma **AND** percentages of each type are unknown/not stated **OR** they are equal percentages<br>• Acinar adenocarcinoma<br>• Clear cell adenocarcinoma<br>• Lepidic adenocarcinoma<br>***Note***: Lepidic adenocarcinoma may or may not have mucinous components.<br>• Micropapillary adenocarcinoma<br>• Papillary adenocarcinoma<br>• Solid adenocarcinoma<br>• Well-differentiated fetal adenocarcinoma<br>***Note:*** This includes a diagnosis of adenocarcinoma **AND** at least two subtypes/variants of adenocarcinoma.<br>2. A combination of histologies **not listed on previous rows** of this table. | Adenocarcinoma with mixed subtypes **8255/3**<br>***Note 1***: 8255 is a "last resort" code.<br>***Note 2:*** See the **Histology Rules** to determine when it is appropriate to use this code for combination histologies other than adenocarcinoma subtypes/variants.<br>***Note 3:*** 8255 does not apply to squamous cell carcinoma NOS and/or subtype/variants of SCC. |

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 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:** Non-small cell lung carcinoma (NSCLC) is a broad group of cancers which includes all **carcinoma types** in Table 3 with the **exception** of:

- Small cell carcinoma/neuroendocrine tumors (NET Tumors) 8041 **AND**
  - All subtypes of small cell 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.

**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.

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Histology Term and Code | Synonym of Specific or NOS | Subtype/variant of NOS and Code |
|---|---|---|
| **Adenocarcinoma 8140**<br><br>**Note 1:** Mucinous adenocarcinoma for lung only is coded as follows:<br>• **8253/3*** when<br>o Behavior unknown/not documented (use staging form to determine behavior when available)<br>o Invasive<br>• **8257/3*** when<br>o Microinvasive<br>o Minimally invasive<br>• **8253/2*** when<br>o Preinvasive<br>o In situ<br><br>**Note 2:** Non-mucinous adenocarcinoma for lung only is coded as follows:<br>• **8256/3*** when<br>o Microinvasive<br>o Minimally invasive<br>• **8250/2*** when<br>o Preinvasive<br>o In situ | Adenocarcinoma NOS<br>Adenocarcinoma in situ **8140/2**<br>Adenocarcinoma invasive **8140/3**<br>Adenocarcinoma, non-mucinous, NOS<br>Minimally invasive adenocarcinoma **8140/3** | Acinar adenocarcinoma/adenocarcinoma, acinar predominant **(for lung only) 8551***<br>Adenoid cystic/adenocystic carcinoma **8200**<br>Colloid adenocarcinoma **8480**<br>Enteric adenocarcinoma/pulmonary intestinal-type adenocarcinoma **8144**<br>Fetal adenocarcinoma **8333**<br>Lepidic adenocarcinoma/adenocarcinoma, lepidic predominant **8250/3***<br>Mucinous carcinoma/adenocarcinoma **(for lung only)**<br>in situ **8253/2*;** invasive **8253/3***<br>minimally invasive **8257/3***<br>microinvasive **8257/3***<br>preinvasive **8253/2***<br>Micropapillary adenocarcinoma/adenocarcinoma, micropapillary predominant **8265**<br>Mixed invasive mucinous and non-mucinous adenocarcinoma **8254***<br>Non-mucinous adenocarcinoma **(for lung only)**<br>in situ **8250/2***<br>microinvasive **8256/3***<br>minimally invasive **8256/3***<br>preinvasive **8250/2***<br>Papillary adenocarcinoma/adenocarcinoma, papillary predominant **8260**<br>Solid adenocarcinoma/adenocarcinoma, solid predominant **8230** |
| **Adenosquamous carcinoma 8560** | | |

**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Histology Term and Code | Synonym of Specific or NOS | Subtype/variant of NOS and Code |
|---|---|---|
| **Epithelial-myoepithelial carcinoma 8562**<br><br>*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*<br>Epimyoepithelial carcinoma<br>Epithelial-myoepithelial tumor of unproven malignant potential*<br>Malignant mixed tumor comprising epithelial and myoepithelial cells<br>Pneumocytic adenomyoepithelioma* | |
| **Epithelioid hemangioepithelioma 9133** | | |
| **Giant cell carcinoma 8031** | | |
| **Intrapulmonary thymoma (arising within lung) 8580/3**<br><br>*Note:* **Intrapulmonary** thymoma is **always malignant** /3. | | |

**Table continues on next page**

**Lung Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Histology Term and Code | Synonym of Specific or NOS | Subtype/variant of NOS and Code |
|---|---|---|
| **Large cell carcinoma 8012**<br><br>***Note 1:*** A diagnosis of large cell carcinoma is usually followed by further diagnostic testing to identify the subtype/variant.<br>***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.<br>***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<br>Large cell carcinoma NOS<br>Large cell carcinoma with no additional stains (subtype/variant – no ICD-O code)<br>Large cell carcinoma with null immunohistochemical features (subtype/variant – no ICD-O code)<br>Large cell carcinoma with unclear immunohistochemical features (subtype/variant – no ICD-O code)<br>Large cell undifferentiated carcinoma | |
| **Large cell neuroendocrine carcinoma 8013**<br>**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** | | |

**Lung Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific or NOS Histology Term and Code | Synonym of Specific or NOS | Subtype/variant of NOS and Code |
|---|---|---|
| **Mucoepidermoid carcinoma 8430**<br><br>***Note:*** Mucoepidermoid tumor **is** listed as a synonym of mucoepidermoid carcinoma in WHO | Mucoepidermoid tumor | |
| **Myoepithelial carcinoma 8982** | | |
| **NUT carcinoma 8023/3***<br>**NUT: nuclear protein in tests NUT/M1 gene rearrangement** | Aggressive t(15:19) positive carcinoma<br>BET-rearranged carcinoma<br>Carcinoma with t(15:19) translocation<br>Midline carcinoma of children and young adults with NUT rearrangement<br>Midline lethal carcinoma<br>NUT midline carcinoma | |
| **PEComa malignant 8714/3**<br><br>***Note:*** Tumor displays perivascular epithelioid (PEC) differentiation | | |

**Lung Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 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**<br><br>*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<br>*Note 2*: Pleomorphic carcinoma has components of **adenocarcinoma** and/or **large cell** carcinoma, also **squamous** carcinoma | | |
| **Sarcoma NOS 8800/3** | | Biphasic synovial sarcoma **9043/3**<br>Epithelioid cell synovial sarcoma **9042/3**<br>Pulmonary artery intimal sarcoma/low-grade malignant myxoid endobronchial tumor **9137/3**<br>Pulmonary myxoid sarcoma with EWSR1 - CREB1 translocation **8842/3**<br>Spindle cell synovial sarcoma **9041/3**<br>Synovial sarcoma **9040/3** |
| **Small cell carcinoma 8041/3**<br><br>*Note 1:* This row applies to neuroendocrine tumors (NET).<br>*Note 2:* Large cell carcinoma with neuroendocrine differentiation lacks NE morphology and is coded as large cell carcinoma, not large cell neuroendocrine carcinoma. | Reserve cell carcinoma<br>Round cell carcinoma<br>SCLC<br>Small cell carcinoma NOS<br>Small cell neuroendocrine carcinoma | Atypical carcinoid **8249/3**<br>Combined small cell carcinoma **8045/3**<br>Typical carcinoid **8240/3**<br>Neuroendocrine carcinoma, NOS<br>Well-differentiated neuroendocrine carcinoma |
| **Spindle cell carcinoma 8032** | | |

**Lung Equivalent Terms and Definitions**
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**(Excludes lymphoma and leukemia M9590 – M9992 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<br>Epidermoid carcinoma NOS<br>Squamous carcinoma<br>Squamous cell carcinoma NOS<br>Squamous cell epithelioma<br>Squamous cell carcinoma in situ **8070/2** | Basaloid carcinoma/basaloid squamous cell carcinoma **8083**<br>Keratinizing squamous cell carcinoma **8071**<br>Non-keratinizing carcinoma **8072** |

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

**Lung Equivalent Terms and Definitions**
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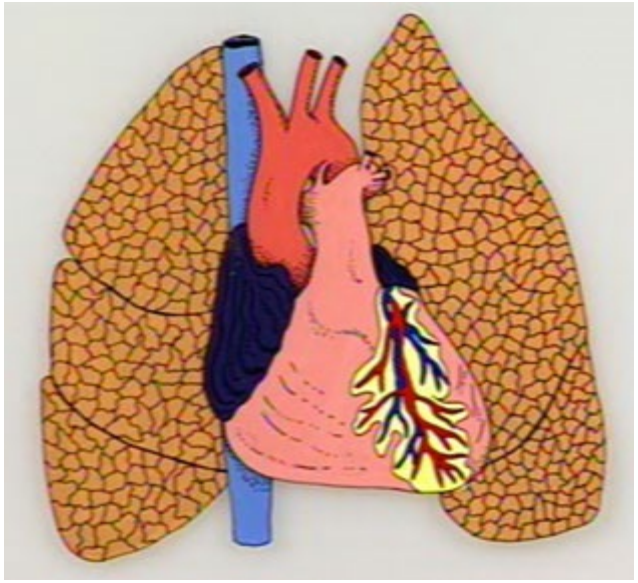
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## Illustrations



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**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
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**Mediastinum**
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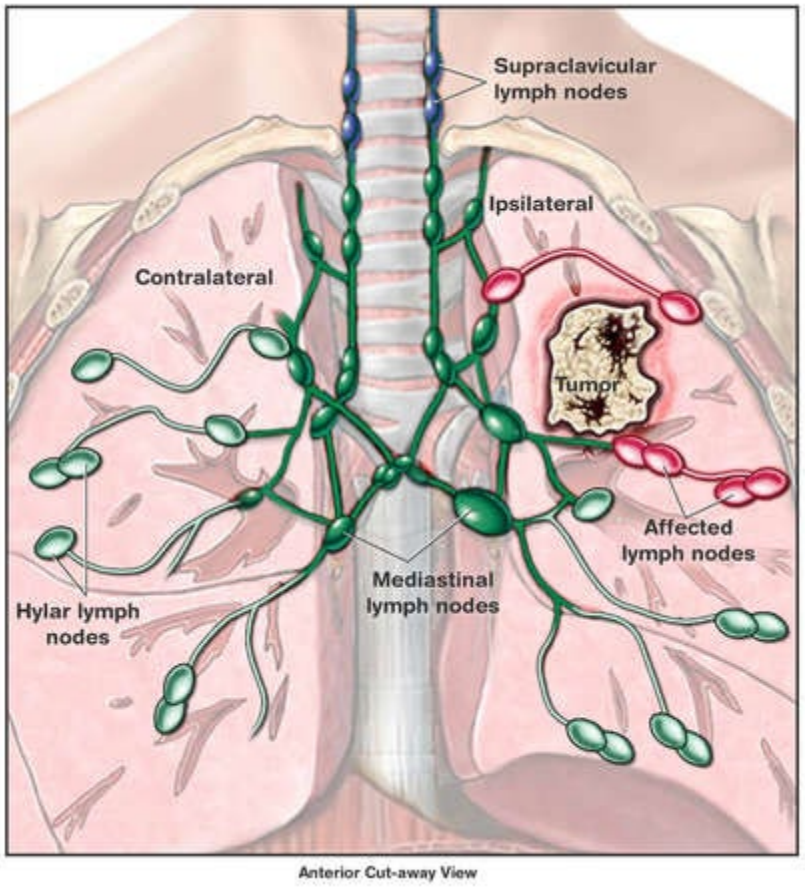

**Lung Equivalent Terms and Definitions**
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**Lymph Nodes Lung**
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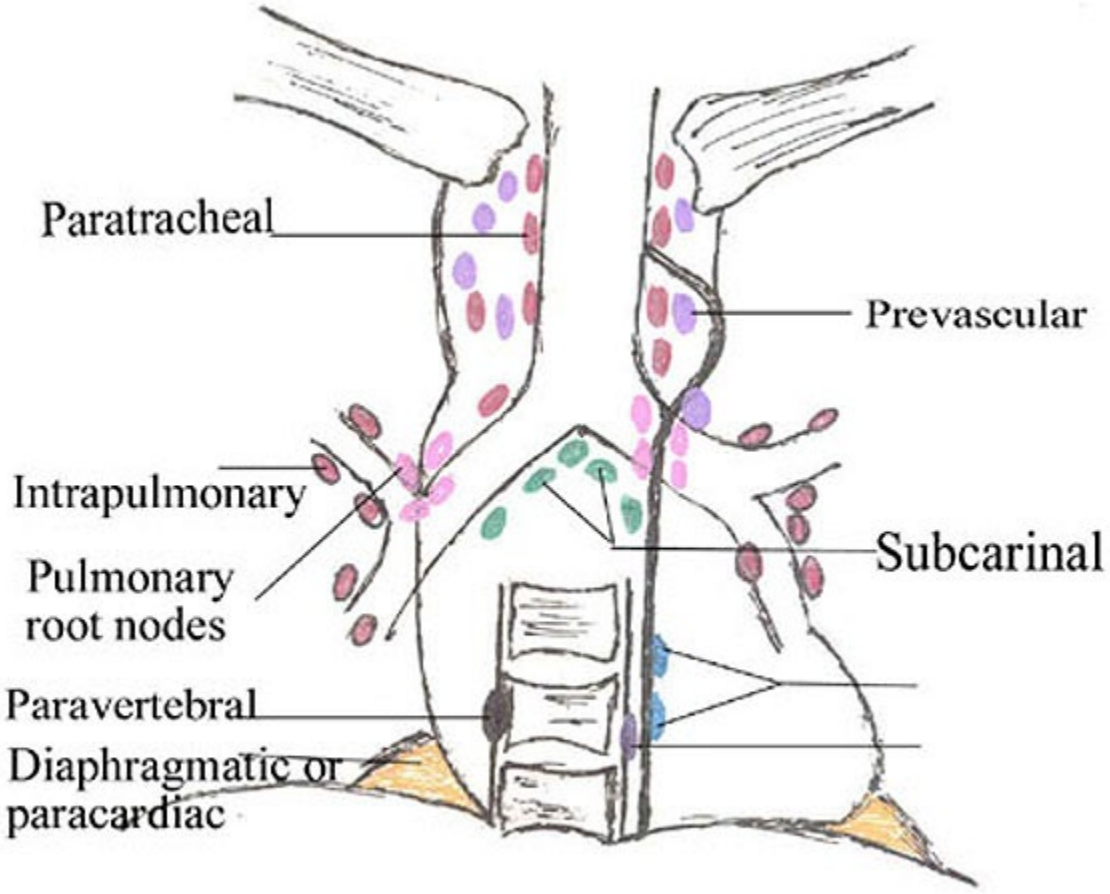
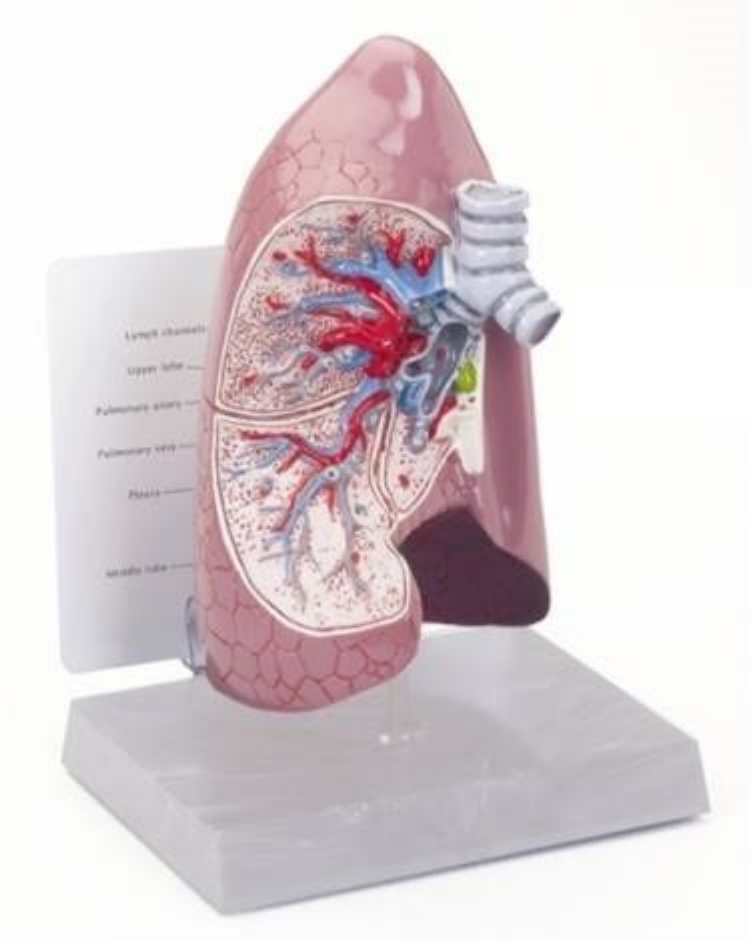


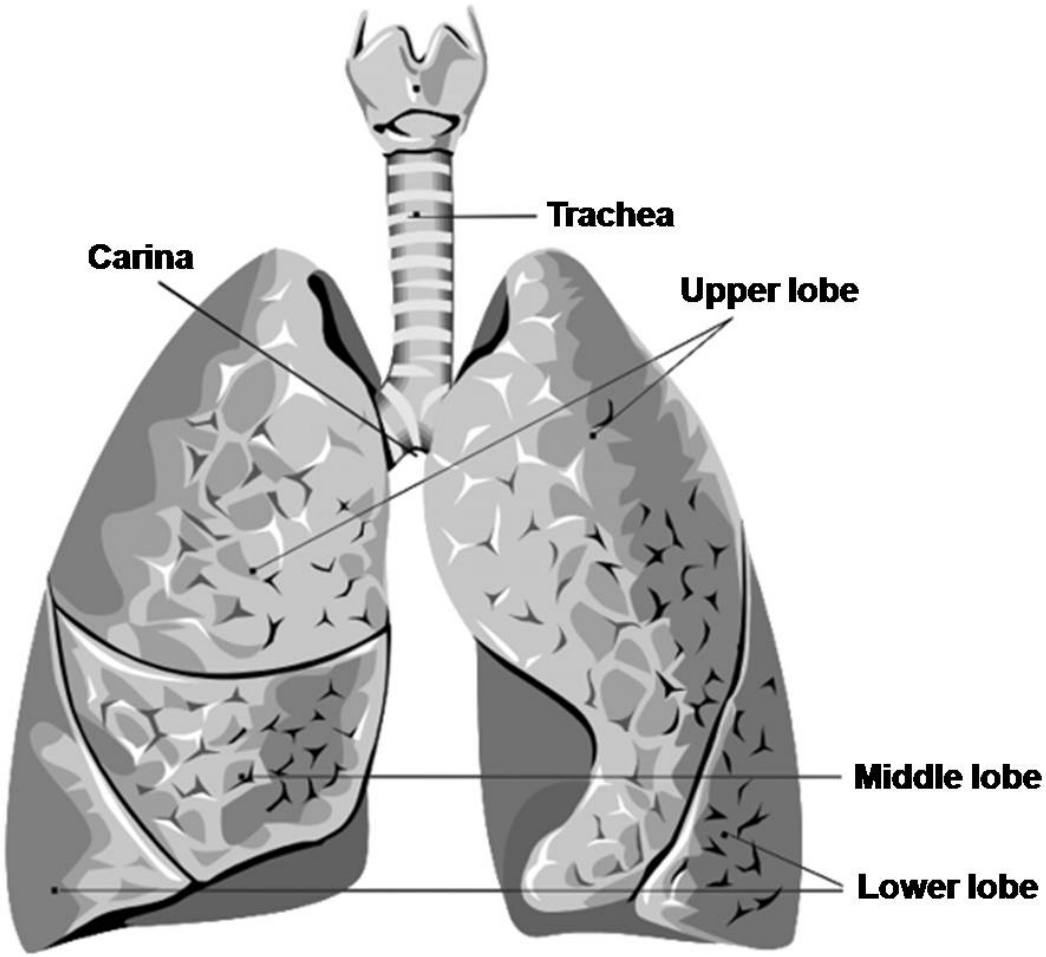
**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

**Inside the Lung**
Used with permission



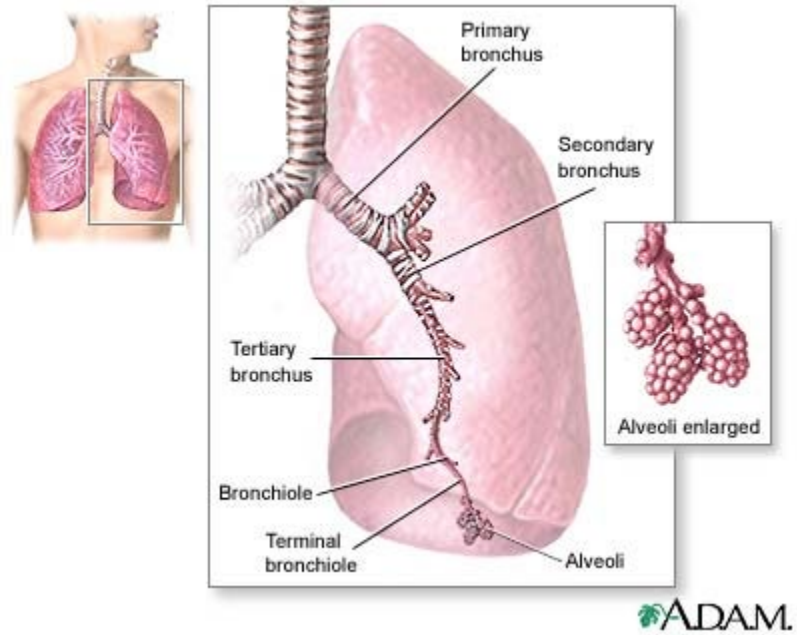
**Lung Equivalent Terms and Definitions**
**C340-C343, C348, C349**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

**Gross Anatomy of Lung**
Used with permission



**End of Mainstem Bronchus; Start of Terminal/Secondary Bronchus**

Used with permission



## Lung Multiple Primary Rules
## C340-C343, C348, C349
## (Excludes lymphoma and leukemia M9590 – M9992 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
# C340-C343, C348, C349
# (Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 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 – M9992 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 (simultaneous or synchronous).

## Lung Multiple Primary Rules
## C340-C343, C348, C349
## (Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 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 – M9992 and Kaposi sarcoma M9140)

## Coding Histology

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

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

   ***Example 1:*** Diagnosis for a single tumor is 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

## 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 – M9992 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
- Small cell neuroendocrine tumors/NET **8041** and a subtype/variant of small cell neuroendocrine tumor/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.

## Lung Histology Rules
## C340-C343, C348, C349
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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.
***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.

**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.**

**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 – M9992 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 which is biopsied 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
- Small cell neuroendocrine tumors/NET **8041** and a subtype/variant of small cell neuroendocrine tumor/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 – M9992 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.

***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.

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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.

## 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.**

**Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions**
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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<br>***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)<br>***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<br>***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<br>***Note:*** Collected as malignant /3 in North America | 1 |
| Pineal parenchymal tumor of intermediate differentiation<br>***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<br>***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 |

## 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

**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 – M9992 and Kaposi sarcoma M9140)**

## 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**<br>Brain stem **C717**<br>Cerebellum NOS **C716**<br>Cerebrum **C710**<br>Frontal lobe **C711**<br>Occipital lobe **C714**<br>Overlapping lesion of brain **C718**<br>Parietal lobe **C713**<br>Temporal lobe **C712**<br>Ventricle NOS **C715** |
| Cranial Nerves | Abducent (cranial nerve VI) **C725**<br>Accessory (cranial nerve XI) **C725**<br>Acoustic (cranial nerve VIII) **C724**<br>Cranial nerve NOS **C725**<br>Facial (cranial nerve VII) C**725**<br>Glossopharyngeal (cranial nerve IX) **C725**<br>Hypoglossal (cranial nerve XII) **C725**<br>Oculomotor (cranial nerve III) **C725**<br>Olfactory (cranial nerve I) **C722**<br>Optic (cranial nerve II) **C723**<br>Trigeminal (cranial nerve V) **C725**<br>Trochlear (cranial nerve IV) **C725**<br>Vagus (cranial nerve X) **C725** |

**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 – M9992 and Kaposi sarcoma M9140)**

| Site Group | Reportable Subsite Terms and Code |
|---|---|
| Ill-Defined Sites Central Nervous System | Nervous system NOS **C729**<br>Overlapping lesion of brain and central nervous system **C728** |
| Intracranial Duct and Glands | Craniopharyngeal duct **C752**<br>Pineal gland **C753**<br>Pituitary gland **C751** |
| Meninges | Cerebral meninges **C700**<br>Meninges NOS **C709**<br>Spinal meninges **C701** |
| Peripheral Nerve and Autonomic Nervous System | Abdomen **C474**<br>Autonomic nervous system NOS **C479**<br>Head, face and neck **C470**<br>Lower limb and hip **C472**<br>Nerves of pelvis **C475**<br>Overlapping lesion of peripheral nerves and autonomic nervous system **C478**<br>Thorax **C473**<br>Trunk NOS **C476**<br>Upper limbs and shoulder **C471**<br>Spinal Nerve NOS **C479** |
| Spinal Sites | Cauda equina/ **C721**<br>Conus medullaris/filum terminale **C720**<br>Meninges NOS **C709**<br>Spinal cord **C720**<br>Spinal meninges **C701** |

**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 – M9992 and Kaposi sarcoma M9140)**

## 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**

**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 – M9992 and Kaposi sarcoma M9140)**

| Specific and NOS Histology Codes | Synonyms | Subtypes/Variants |
|---|---|---|
| Anaplastic ganglioglioma **9505** | | |
| Astroblastoma **9430** | | |
| Astrocytoma NOS **9400** | Diffuse astrocytoma IDH-mutant<br>Diffuse astrocytoma IDH-wildtype<br>Diffuse astrocytoma NOS | Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401**<br>Gemistocytic astrocytoma IDH-mutant **9411**<br>Pleomorphic xanthroastrocytoma /anaplastic pleomorphic xanthroastrocytoma **9424** |
| Choriocarcinoma **9100** | | |
| Choroid plexus carcinoma **9390** | | |
| CNS embryonal tumor with rhabdoid features **9508** | Atypical teratoid/rhabdoid tumor<br>Embryonal tumor with rhabdoid features | |
| CNS ganglioneuroblastoma **9490** | | CNS embryonal tumor **9473** |
| CNS neuroblastoma **9500** | | |
| Diffuse midline glioma H3 K27M mutant **9385*** | | |
| Embryonal carcinoma **9070** | | Yolk sac tumor **9071** |
| Embryonal tumor with multilayered rosettes C19MC-altered **9478*** | Embryonal tumor with multilayered rosettes, NOS<br>ETMR | |
| Ependymoma **9391** | Clear cell ependymoma<br>Tanycytic ependymoma | Anaplastic ependymoma **9392**<br>Ependymoma, RELA fusion-positive **9396***<br>Papillary ependymoma **9393** |
| Epithelioid hemangioendothelioma **9133** | | |
| Germinoma **9064** | | |

**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 – M9992 and Kaposi sarcoma M9140)**

| Specific and NOS Histology Codes | Synonyms | Subtypes/Variants |
|---|---|---|
| Glioblastoma NOS **9440** | Glioblastoma multiforme<br>GBM<br>Glioblastoma, IDH wild-type<br>Epithelioid glioblastoma | Giant cell glioblastoma **9441**<br>Glioblastoma IDH-mutant **9445***<br>Gliosarcoma **9442** |
| Immature teratoma **9080** | | Mixed germ cell tumor **9085**<br>Teratoma with malignant transformation **9084** |
| Malignant meningioma **9530** | Anaplastic meningioma | Papillary/rhabdoid meningioma **9538** |
| Malignant peripheral nerve sheath tumor **9540** | Malignant perineurioma<br>MPNST<br>MPNST with perineural differentiation | Epithelioid malignant peripheral nerve sheath tumor **9542** |
| Medulloblastoma NOS **9470** | Classic medulloblastoma | Anaplastic/large cell medulloblastoma **9474**<br>Medulloblastoma described as one of the following **9471**<br>Desmoplastic<br>SHH-activated and TP53-wildtype<br>With extensive nodularity<br>Nodular<br>Medulloblastoma non-WNT/non-SHH; medulloblastoma group 3 or group 4 **9477***<br>Medulloblastoma SHH-activated and TP53-mutant **9476***<br>Medulloblastoma WNT-activated **9475*** |
| Medulloepithelioma **9501** | | |
| Meningeal melanoma **8720** | | Meningeal melanomatosis **8728** |
| Oligoastrocytoma NOS **9382** | Anaplastic oligoastrocytoma NOS | |
| Oligodendroglioma NOS **9450** | Oligodendroglioma 1p/19q-codeleted<br>Oligodendroglioma IDH-mutant | Anaplastic oligodendroglioma NOS **9451**<br>IDH-mutant |

**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 – M9992 and Kaposi sarcoma M9140)**

| Specific and NOS Histology Codes | Synonyms | Subtypes/Variants |
|---|---|---|
| ***Note:*** Oligodendroglioma NOS is used when molecular markers cannot fully be determined | Oligodendroglioma IDH-mutant and 1p/19q-codeleted | 1p/19q-codeleted<br>IDH-mutant and 1p/19q-codeleted |
| Peripheral primitive neuroectodermal tumor **9364** | Ewing sarcoma<br>pPNET | |
| Pilocytic astrocytoma **9421**<br><br>***Note:*** ICD-O-3 lists a behavior code of /1. It is collected as /3 in North America. | | Pilomyxoid astrocytoma **9425** |
| Pineal parenchymal tumor of intermediate differentiation **9362** | Pineoblastoma | Papillary tumor of the pineal region **9395** |
| Sarcoma NOS **8800**<br><br>***Note 1:*** **Chondrosarcoma 9220** has the following subtype/variant:<br>Mesenchymal chondrosarcoma **9240**<br><br>***Note 2:*** **Leiomyosarcoma 8890** has the following subtypes/variants:<br>Epithelioid leiomyosarcoma **8891**<br>Myxoid leiomyosarcoma **8896** | | Angiosarcoma **9120**<br>Chondrosarcoma **9220**<br>Mesenchymal chondrosarcoma **9240**<br>Leiomyosarcoma/granular cell leiomyosarcoma/inflammatory leiomyosarcoma **8890**<br>Epithelioid leiomyosarcoma **8891**<br>Myxoid leiomyosarcoma **8896**<br>Osteosarcoma **9180**<br>Undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma **8802** |
| Solitary fibrous tumor grade 3 **8815** | Hemangiopericytoma grade 3<br>Solitary fibrous tumor/Hemangiopericytoma grade 3 (CNS) | |

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

**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 – M9992 and Kaposi sarcoma M9140)**

## 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** |

| 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<br><br>***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.<br><br>Superior orbital fissure.<br><br>The foramen rotundum<br><br>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**<br>. | • **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**<br>• **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**<br>• **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 – M9992 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**<br><br>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<br>Between the i**nternal jugular** vein and **internal carotid** artery<br>Lies on the **stylopharyngeus** and middle **pharyngeal** constrictor muscle<br>Passes under the **hypoglossus** muscle<br>Palatine tonsil<br>Extends to mucous **glands** of the **mouth**, and **base** of the **tongue C470** |

**Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions**
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| 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**.<br>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**<br>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**<br>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.

## 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**
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**(Excludes lymphoma and leukemia M9590 – M9992 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**

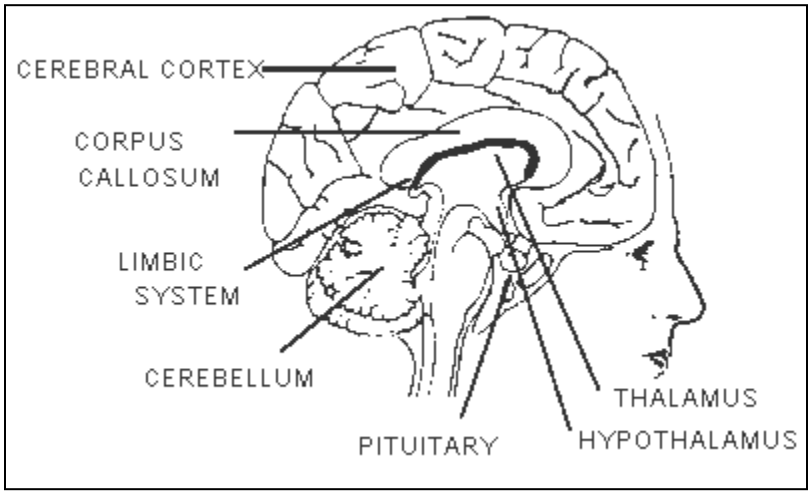
The table identifies non-malignant tumors that have the **potential** of **transforming to** a malignant tumor (**new primary**).

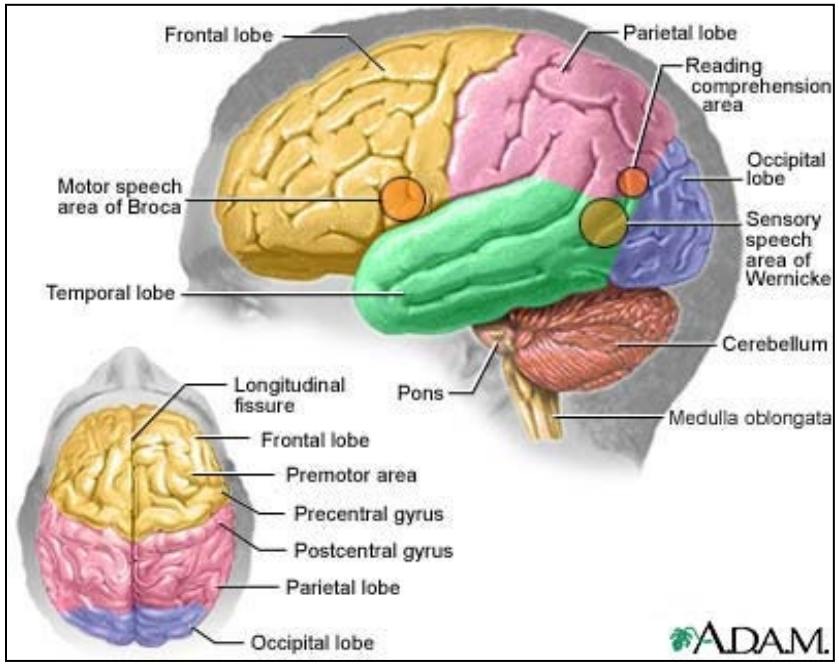
Use **Table 6** when directed to by the Multiple Primary Rules.

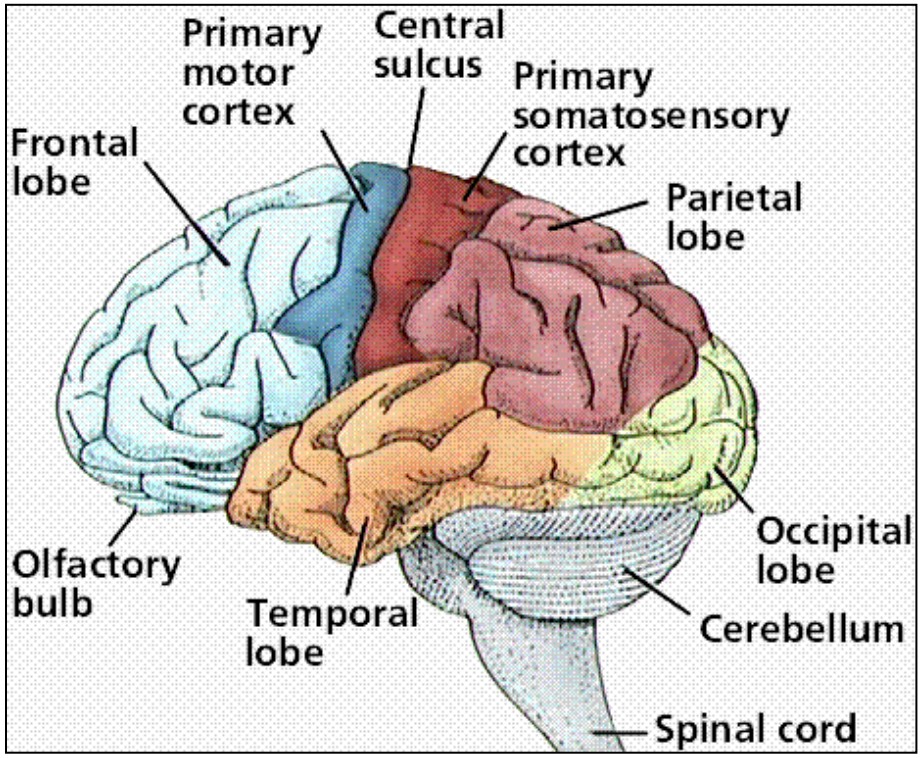
**Column 1** is the **non-malignant** ICD-O histology term and code.
**Column 2** is the **malignant** /3 ICD-O histology term and code to which the non-malignant tumor can transform.

| 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** |

## Illustrations







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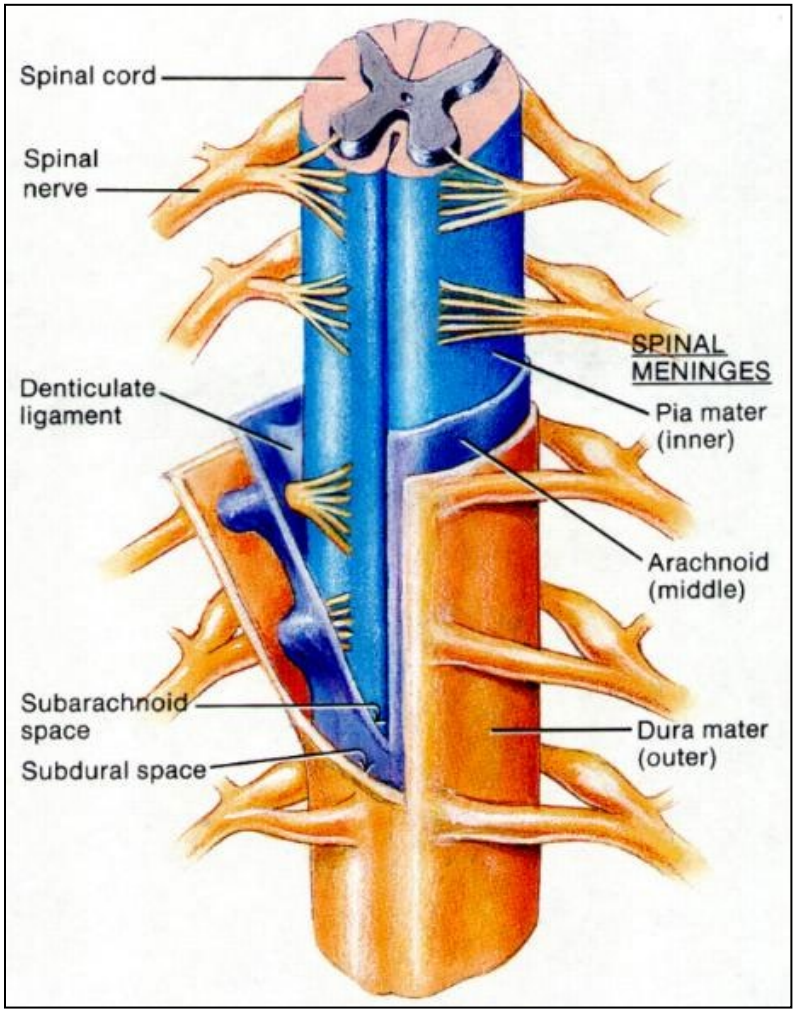

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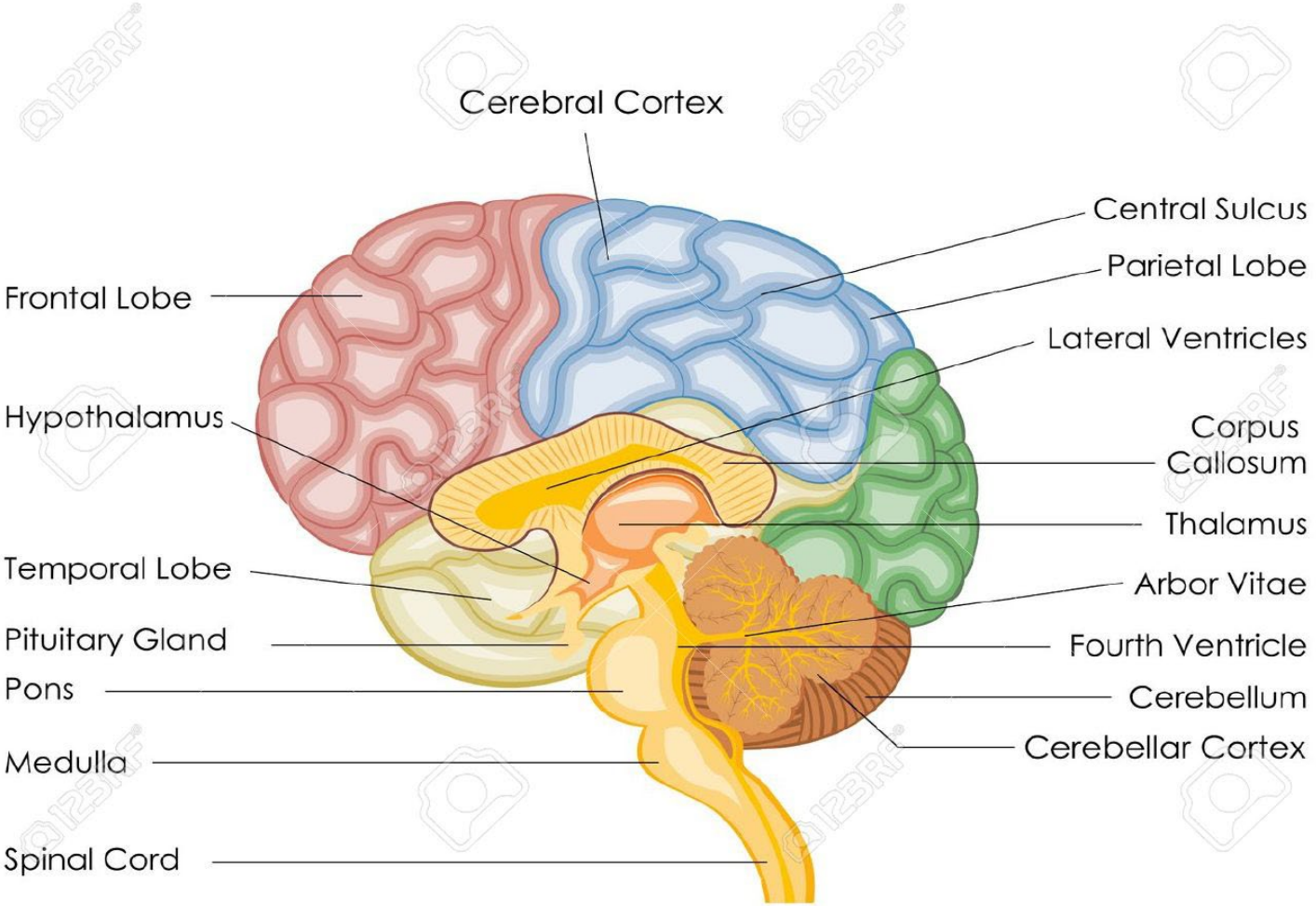

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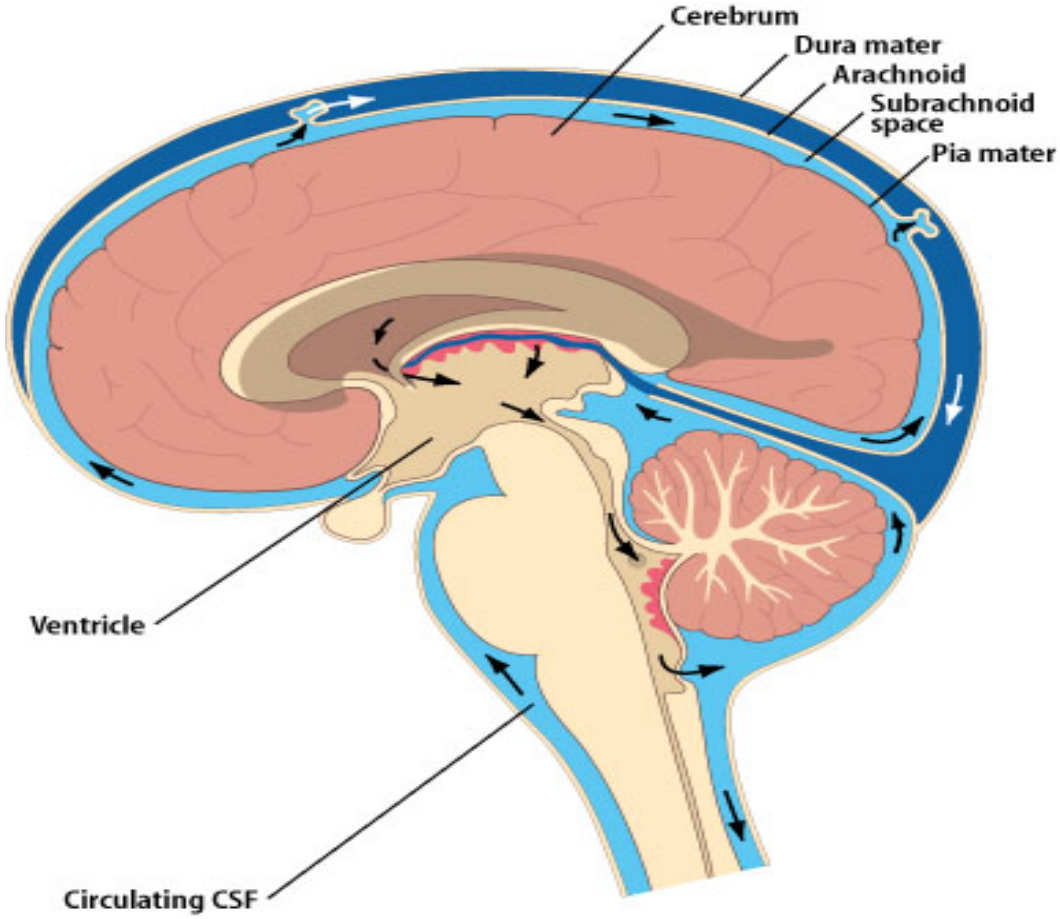

## Malignant CNS and Peripheral Nerves Multiple Primary Rules
## C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

***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 – M9992 and Kaposi sarcoma M9140)

### Single Tumor

**IMPORTANT:** The **major difference** between **M4** and **M5** is:
**M4: 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 **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** Code 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] (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:*** 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 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 (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 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 – M9992 and Kaposi sarcoma M9140)

### Multiple Tumors

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

- In the CNS (see **Table 2**) AND in a site other than the CNS
  ***Example***: Patient has a 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 M8)

**IMPORTANT:** The **major difference** between **M4** and **M5** is:
**M4: 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 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 – M9992 and Kaposi sarcoma M9140)

**Rule M6** 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**
- 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 – M9992 and Kaposi sarcoma M9140)

**Rule M7** 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 – M9992 and Kaposi sarcoma M9140)

**Rule M8** 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 M9** 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 M10** 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 – M9992 and Kaposi sarcoma M9140)

**Rule M11** 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:*** Each row in the table is a **distinctly different** histology.

**Rule M12** 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 – M9992 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 – M9992 and Kaposi sarcoma M9140)**

## Coding Histology

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

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

   ***Example 1:*** Diagnosis for a single tumor is 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 – M9992 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.

**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:*** 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 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 – M9992 and Kaposi sarcoma M9140)

### Introduction

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

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

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

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

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

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

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

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

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

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

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## Equivalent or Equal Terms

These terms can be used interchangeably:

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

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## Terms that are NOT Equivalent or Equal

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

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

## Changes from 2007 MPH Rules

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

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

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

## Reportability Criteria for Non-Malignant CNS Neoplasms

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

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

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## Directory of Sections and Tables

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

**Non-Malignant CNS Equivalent Terms and Definitions**
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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<br>***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)<br>***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<br>***Note:*** Tissue/pathology report or CAP will specify whether it is WHO Grade 2 or 3 | 2 or 3 |
| Perineuroma | 1 |
| Pilocytic astrocytoma<br>***Note:*** ICD-O-3 lists a behavior code of /1. US and Canada collect as /3. | 1 |
| Pineal parenchymal tumor of intermediate differentiation<br>***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<br>***Note:*** Tissue/pathology will specify WHO Grade 1, 2, or 3 | 1, 2, or 3 |
| Spindle cell oncocytoma | 1 |
| Subependymal giant cell astrocytoma | 1 |
| Subependymoma | 1 |

## Section 2: Reportable Primary Sites and Histologies

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

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

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

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

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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**<br>Brain stem **C717**<br>Cerebellum NOS **C716**<br>Cerebrum **C710**<br>Frontal lobe **C711**<br>Occipital lobe **C714**<br>Overlapping lesion of brain and central nervous system **C718**<br>Parietal lobe **C713**<br>Temporal lobe **C712**<br>Ventricle NOS **C715** |
| Cranial Nerves | Abducent (cranial nerve VI) **C725**<br>Accessory (cranial nerve XI) **C725**<br>Acoustic (cranial nerve VIII) **C724**<br>Cranial nerve NOS **C725**<br>Facial (cranial nerve VII) C**725**<br>Glossopharyngeal (cranial nerve IX) **C725**<br>Hypoglossal (cranial nerve XII) **C725**<br>Oculomotor (cranial nerve III) **C725**<br>Olfactory (cranial nerve I **C722**)<br>Optic (cranial nerve II) **C723**<br>Trigeminal (cranial nerve V) **C725**<br>Trochlear (cranial nerve IV) **C725**<br>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**<br>Overlapping lesion of brain and central nervous system **C728** |
| Intracranial Duct and Glands | Craniopharyngeal duct **C752**<br>Pineal gland **C753**<br>Pituitary gland **C751** |
| Meninges | Cerebral meninges **C700**<br>Meninges NOS **C709**<br>Spinal meninges **C701** |
| Spinal Sites | Cauda equina/ **C721**<br>Conus medullaris/filum terminale **C720**<br>Meninges NOS **C709**<br>Spinal cord **C720**<br>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<br>**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<br>**C725**<br><br>***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**<br>• **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.<br>• **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<br>**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<br>**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<br>**C724** | Internal acoustic meatus | Originates in the **brain stem** (**medulla oblongata**) **between** the base of the brain (**pons**) and the **spinal cord**<br><br>Both the **vestibular** branch and the **cochlear** branch are located in the **inner ear** | |
| Glossopharyngeal CN 9<br>**C725** | Jugular foramen | **Originates** in the anterior portion of the **medulla oblongata** | **Jugular foramen**<br>Between the i**nternal jugular** vein and **internal carotid artery**<br>Lies on the **stylopharyngeus** and **middle pharyngeal constrictor muscle**<br>Passes under the **hypoglossus muscle**<br>**Palatine tonsil**<br>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<br>**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<br>**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**.<br><br>The fibers of the spinal accessory nerve coalesce to **form spinal rootlets, roots**, and finally the **spinal accessory nerv**e itself.<br><br>The nerve **exits** the skull through the jugular foramen. It then runs along the internal **carotid artery** within the **neck** and reaches the **sternocleidomastoid** muscle and the **trapezius.** |

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

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

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

## Table 4: Non-Reportable Neoplasms

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

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

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

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

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

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

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

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

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

Use **Table 5** to:

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

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

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

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

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

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

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

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

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

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

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

| NOS/Specific Histology Term and Code | Synonyms | Subtypes/Variants Histology Term and Codes |
|---|---|---|
| **Angiocentric glioma 9431/1*** | Angiocentric neuroepithelial tumor<br>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**<br>Papillary craniopharyngioma **9352/1** |
| **Desmoplastic infantile astrocytoma and ganglioglioma 9412/1** | | |

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

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

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

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

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

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

## Section 3: Additional Information to Complete Abstract

### Conflicting Information on Pathology Report(s)

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

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

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

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

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

## Table 7: Paired Sites

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

| Paired Sites and Codes |
|---|
| Acoustic nerve **C724** |
| Cerebral meninges **C700** |
| Cerebrum **C710** |
| Cranial nerves **C725** |
| Frontal lobe **C711** |
| Occipital lobe **C714** |
| Olfactory nerve **C722** |
| Optic nerve **C723** |
| Parietal lobe **C713** |
| Temporal lobe **C712** |

***Note 1:*** Midline tumors are most common for the following histologies: glioblastoma multiforme, meningiomas, lymphomas (usually located near the midline), and epidermoid cysts (which can cross the midline via the subarachnoid space).
***Note 2:*** SEER allows laterality to be coded for sites other than those in the table.

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

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

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

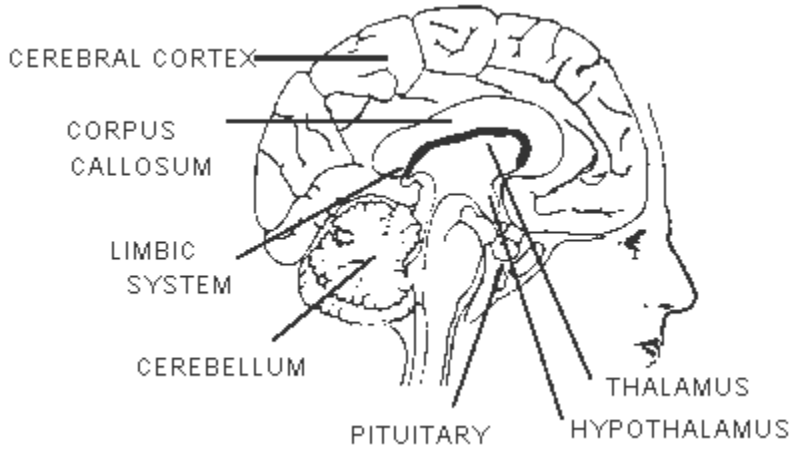
- Residual tumor becomes more aggressive /3 **OR**
- The tumor **recurs** as a **more aggressive /3 histology**

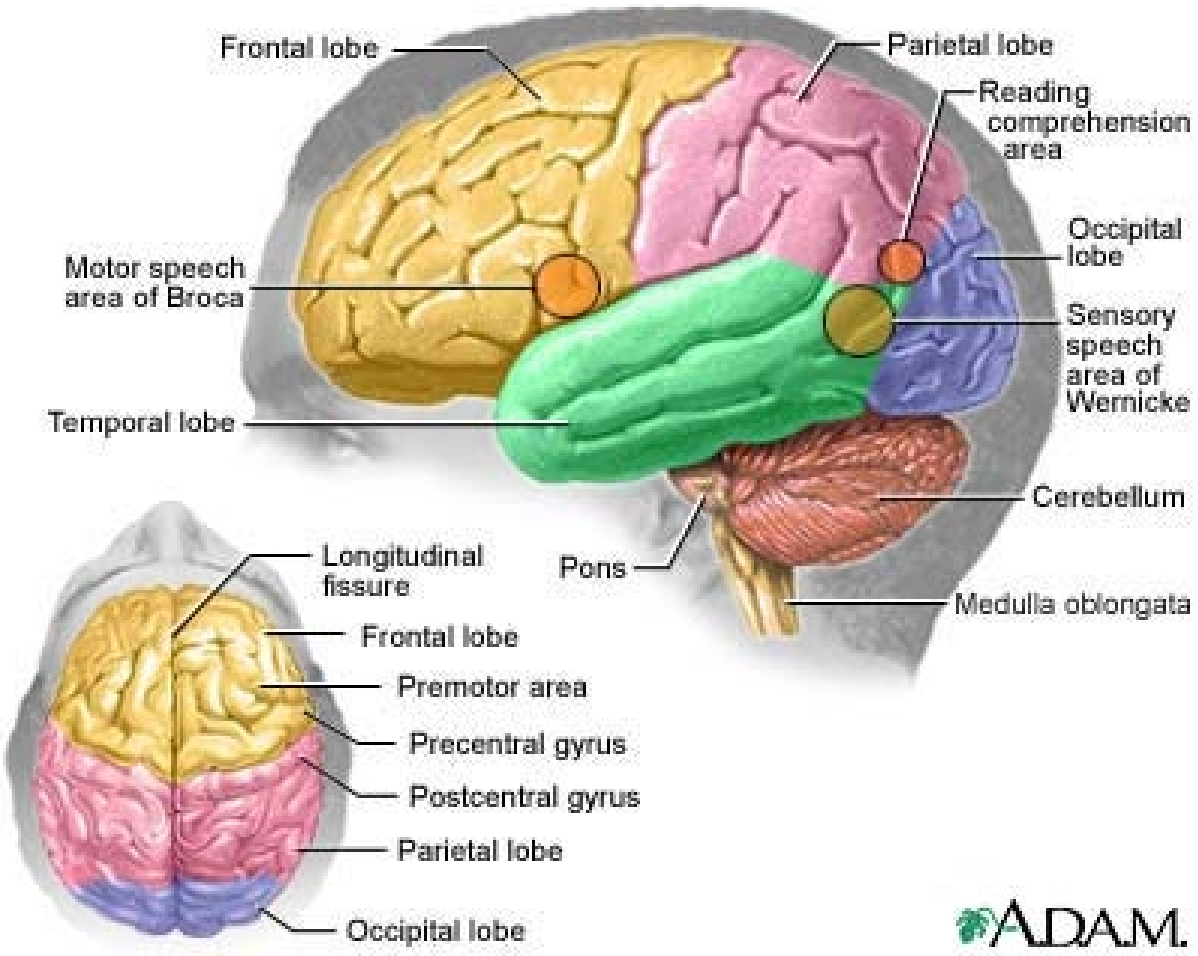
The table identifies non-malignant tumors that have the **potential** of **transforming to** a malignant tumor (**new primary**).

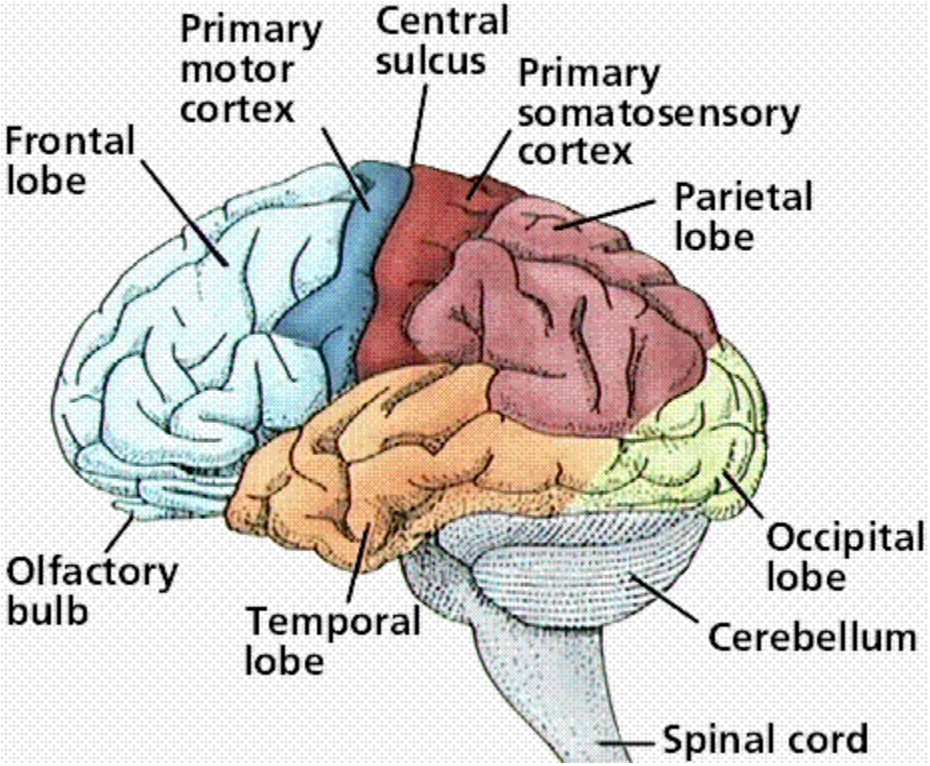
**Column 1** is the **non-malignant** ICD-O histology term and code.
**Column 2** is the **malignant** /3 ICD-O histology term and code to which the non-malignant tumor can transform.

| Original Histology and Code | Transformed Histology and Code |
|---|---|
| Chondroma **9220/0** | Chondrosarcoma **9220/3** |
| Ganglioglioma **9505/1** | Anaplastic ganglioglioma **9505/3** |
| Hemangioma **9120/0** | Angiosarcoma **9120/3** |
| Hemangiopericytoma **9150/1** | Anaplastic hemangiopericytoma **9150/3** |
| Leiomyoma **8890/0** | Leiomyosarcoma **8890/3** |
| Lipoma **8850/0** | Liposarcoma **8850/3** |
| Osteoma **9180/0** | Osteosarcoma **9180/3** |
| Perineurioma **9571/0** | Malignant perineurioma **9571/3** |
| Rhabdomyoma **8900/0** | Rhabdomyosarcoma **8900/3** |
| Teratoma **9080/1** | Immature teratoma **9080/3** |
| Teratoma, mature **9080/0** | Immature teratoma **9080/3** |

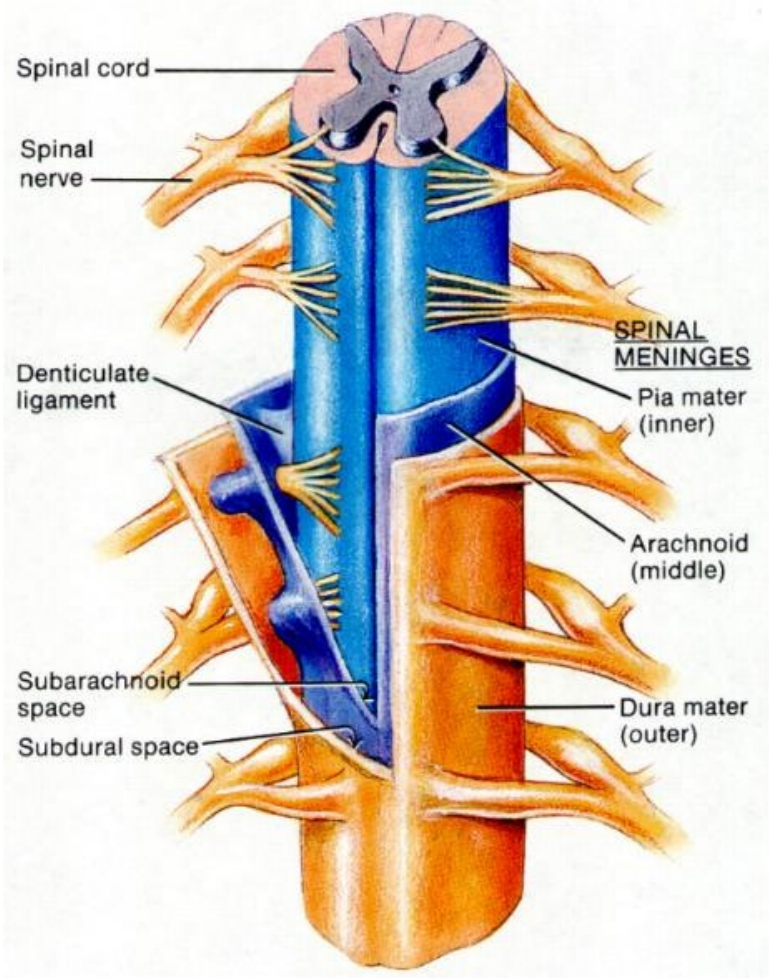
## Illustrations



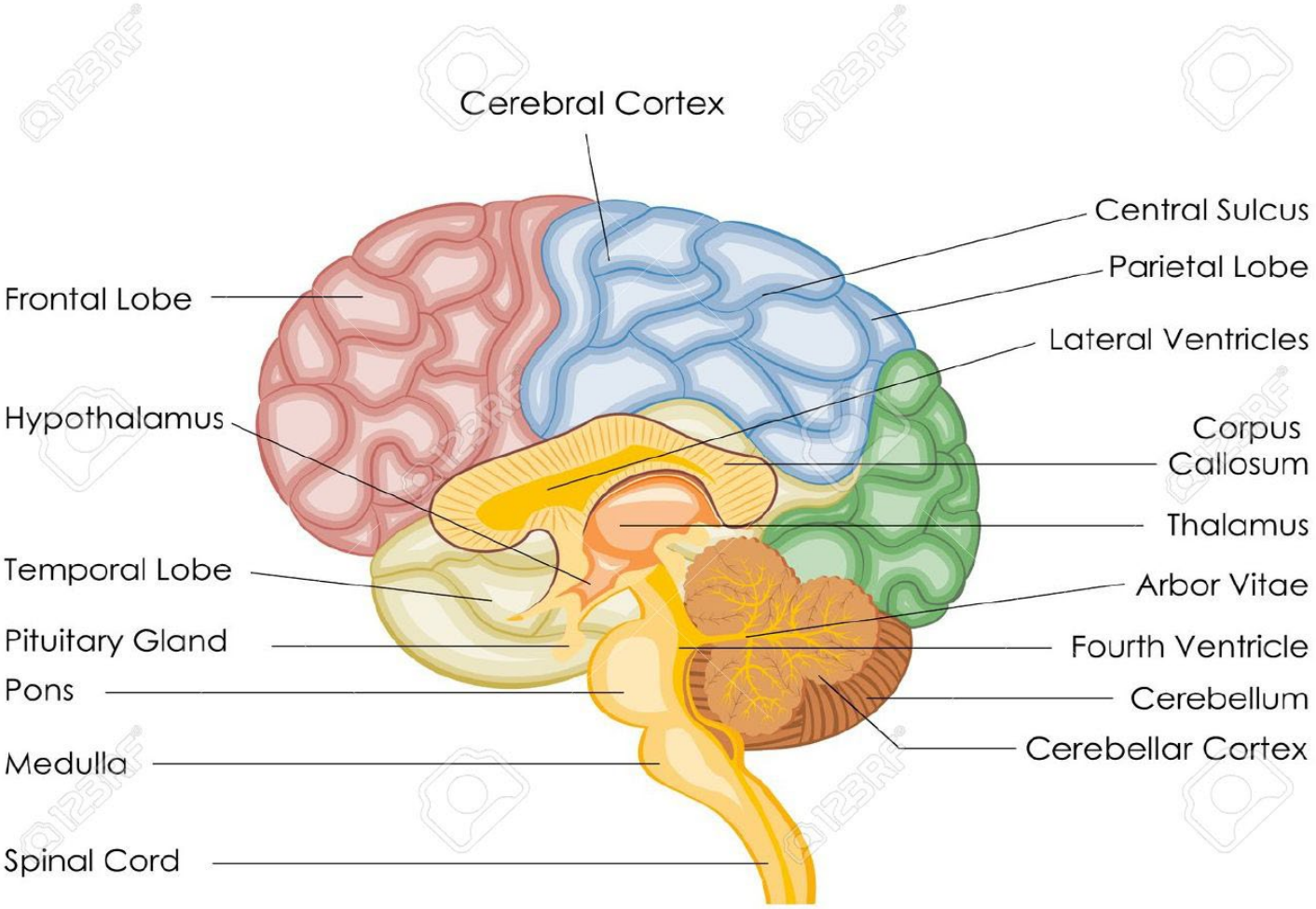




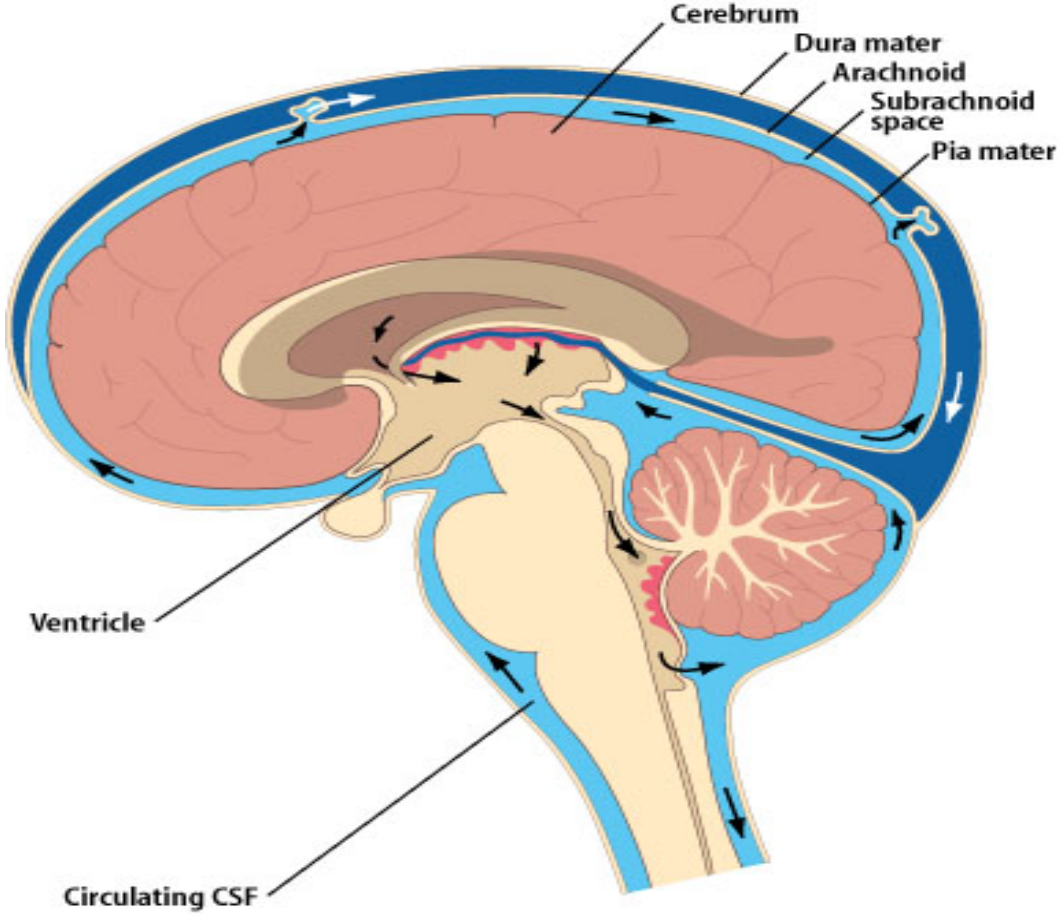
**Non-Malignant CNS Equivalent Terms and Definitions**
**C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**



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



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



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

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

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

### Unknown if Single or Multiple Tumors

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

***Note 1:*** Use this rule only after all information sources have been exhausted
***Note 2:*** Examples of cases with minimal information include

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

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

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

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

### Single Tumor

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

**Rule M2** Abstract a **single primary**[i] when there is a **single tumor.**

***Note 1:*** A single tumor is **always** a single primary.
***Note 2:*** The tumor may overlap onto or extend into an adjacent/contiguous site or subsite.
***Note 3:*** The tumor may have multiple histologic components.
***Note 4:*** The tumor may be diagnosed as borderline /1 on scans and subsequent pathology proves a benign /0 tumor.
***Example:*** Patient has a MRI with a diagnosis of a benign /0 tumor. The pathology report from the subsequent surgery shows a borderline /1 tumor. This is a single tumor and **a single primary**. (It is the same tumor as seen on the MRI; better information is provided by the pathology report.)

**Rule M3** Abstract a **single primary**[i] (the malignant) when a single tumor meets the following two criteria:

1. The original diagnosis was non-malignant /0 or /1 **AND**
   - First course treatment was active surveillance (no tumor resection). Diagnosis was:
     - Clinical
     - Radiographic
     - Stereotactic biopsy

2. Subsequent resection pathology is malignant /3

***Note 1:*** This is a **new rule** which **clarifies** that a single tumor is **always** a single primary and the malignant behavior is reported.
***Note 2:*** Use the Malignant CNS and Peripheral Nerves Rules to code histology.
***Note 3:*** The **resection pathology** is **more accurate** than clinical, radiographic or stereotactic biopsy information. While stereotactic biopsy provides a pathologic specimen, it is small and may not have included the malignant portion of tumor.
***Note 4:*** There is **no time requirement** from initial diagnosis to resection.
***Note 5:*** Edit the original abstract as follows:
- Do **not** change **date of diagnosis**.
- For cases which have been abstracted, **change behavior** code on original abstract from /0 or /1 to /3.
- **Report** all data changes for cases which have been submitted to the central registry.

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

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

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

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

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

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

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

***Note 2:*** This is a single tumor; single primary

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

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

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

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

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

**This is the end of instructions for Single Tumor.**

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

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

### Multiple Tumors

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

**Rule M10** Abstract a **single primary**[i] when there are separate/non-contiguous tumors in the brain (multicentric/multifocal) with the **same histology XXXX**. Tumors may be in any of the following locations and/or lateralities:

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

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

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

***Note:*** The same row means the tumors are:

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

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

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

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

**This is the end of instructions for Multiple Tumors**

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

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

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

## Priority Order for Using Documentation to Identify Histology

**IMPORTANT NOTES**

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

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

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

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

**This is a hierarchical list of source documentation:**

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

- Biomarkers are not listed because they change rapidly.

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

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

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

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

***Note 4:*** The CAP protocol is a checklist which

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

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

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

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

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

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

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

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

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

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

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

## Coding Histology

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

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

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

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

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

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

2. Code the histology described as **differentiation** or **features/features of <u>ONLY</u>** when there is a specific ICD-O code for the "NOS with ____ features" or "NOS with ____ differentiation".
   ***Note***: Do not code differentiation or features when there is no specific ICD-O code.

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

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

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

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

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

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

List of Ambiguous Terminology

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

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

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

### Single Tumor

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

**This is the end of instructions for Single Tumor.**

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

## Multiple Tumors Abstracted as a Single Primary

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

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

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

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

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

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

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

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

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

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

***Note 1:*** Only report tumors such as:

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

## Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
## C659, C669, C670-C679, C680-C689
## (Excludes lymphoma and leukemia M9590 – M9992 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 – M9992 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 – M9992 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.
- Carcinoma; adenocarcinoma
- 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
## C659, C669, C670-C679, C680-C689
## (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

### 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**
- **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<br>Vault<br>Vertex |
| Bladder, lateral wall **C672** | Lateral to ureteral orifice<br>Left wall<br>Right wall<br>Sidewall |
| Bladder neck **C675** | Internal urethral orifice<br>Vesical neck |
| Bladder NOS **C679** | Lateral posterior wall (**no hyphen**) |
| Bladder, overlapping lesion **C678** | Fundus<br>Lateral-posterior wall (**hyphen**) |
| Bladder, posterior wall **C674** | - |

**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Site Term and code | Synonyms |
|---|---|
| Bladder, trigone **C670** | Base of bladder<br>Below interureteric crest<br>Below interureteric field<br>Below interureteric ridge<br>Floor of bladder |
| Bladder, urachus **C677** | Mid umbilical ligament<br>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<br>Pelviureteric junction<br>Renal calyces<br>Renal calyx |
| Ureter **C669** | - |
| Urethra **C680** | Cowper gland<br>Prostatic utricle<br>Urethral gland |
| Urinary system NOS **C689** | - |

**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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**

**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

| Specific and NOS Histology Codes | Synonyms | Subtypes/Variants |
|---|---|---|
| Adenocarcinoma NOS **8140**<br><br>*Note*: Urachal **carcinoma** NOS is coded 8010/3. Urachal **adenocarcinoma** is coded 8140/3. | Mixed adenocarcinoma<br>Urachal adenocarcinoma | Clear cell carcinoma **8310**<br>Endometrioid carcinoma **8380**<br>Enteric adenocarcinoma **8144**<br>Mucinous adenocarcinoma **8480** |
| Malignant melanoma **8720/3** | | |
| Malignant perivascular epithelioid cell tumor **8714/3** | Malignant PEComa | |
| Sarcoma NOS **8800/3**<br><br>***Note:*** Rhabdomyosarcoma **8900** is a NOS with a subtype/variant of embryonal rhabdomyosarcoma/sarcoma botryoides **8910/3.** | | Angiosarcoma **9120/3**<br>Chondrosarcoma **9220/3**<br>Leiomyosarcoma **8890/3**<br>Liposarcoma **8850/3**<br>Malignant peripheral nerve sheath tumor (MPNST) **9540/3**<br>Pleomorphic sarcoma **8802/3**<br>Rhabdomyosarcoma **8900/3**<br>Embryonal rhabdomyosarcoma/sarcoma botryoides **8910/3** |
| Small cell neuroendocrine carcinoma **8041** | Neuroendocrine carcinoma<br>SmCC | Large cell neuroendocrine tumor **8013**<br>Well-differentiated neuroendocrine tumor **8240** |
| Squamous cell carcinoma **8070** | Pure squamous cell carcinoma<br>SCC | Verrucous carcinoma **8051** |

**Table continues on next page**

**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

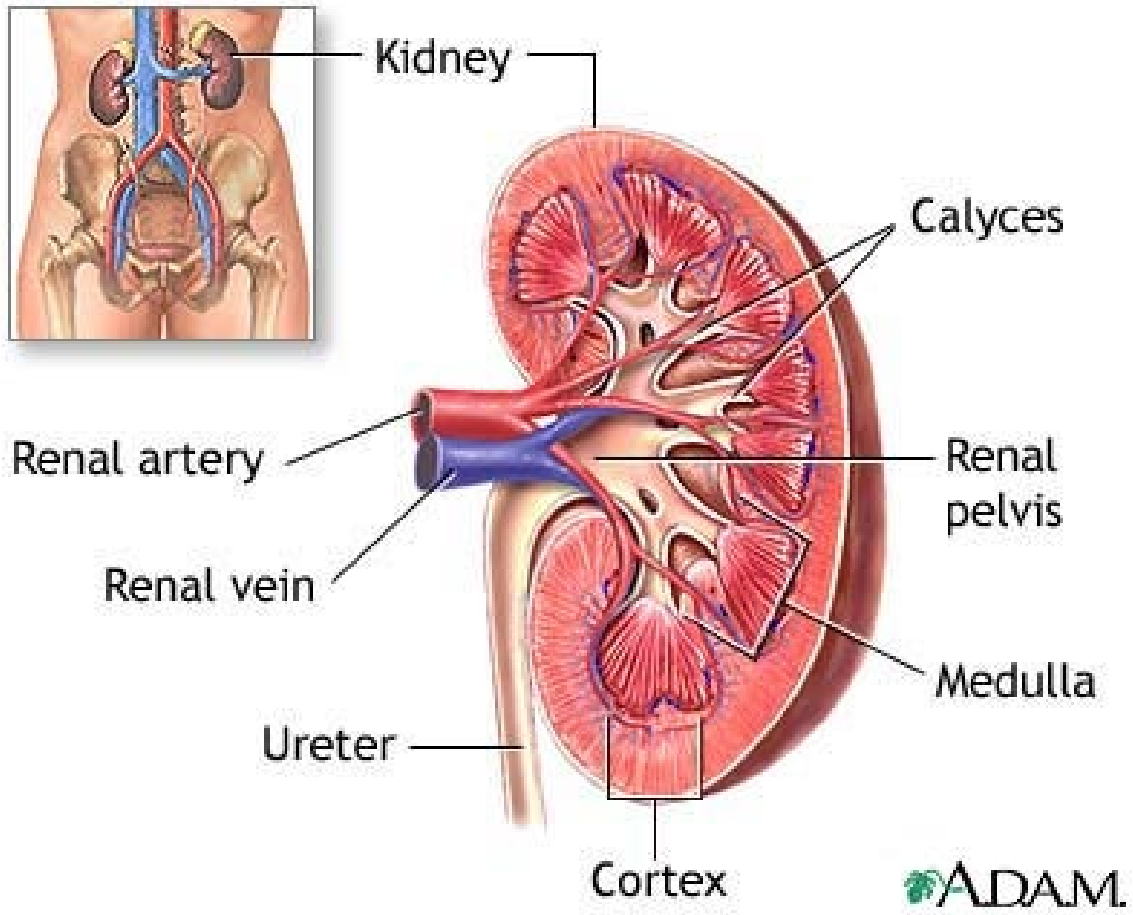
| Specific and NOS Histology Codes | Synonyms | Subtypes/Variants |
|---|---|---|
| Urothelial carcinoma **8120**<br><br>***Note 1:*** Previously called **transitional cell** carcinoma, a term that is no longer recommended.<br>***Note 2:*** Micropapillary **8131** is a subtype/variant of papillary urothelial carcinoma **8130**. It is an invasive /3 neoplasm with aggressive behavior. | Clear cell (glycogen-rich) urothelial carcinoma **8120/3**<br>Infiltrating urothelial carcinoma **8120/3**<br>Infiltrating urothelial carcinoma with divergent differentiation **8120/3**<br>Infiltrating urothelial carcinoma with endodermal sinus lines **8120/3**<br>Infiltrating urothelial carcinoma with glandular differentiation **8120/3**<br>Infiltrating urothelial carcinoma with squamous differentiation **8120/3**<br>Infiltrating urothelial carcinoma with trophoblastic differentiation **8120/3**<br>Lipid-rich urothelial carcinoma **8120/3**<br>Microcystic urothelial carcinoma **8120/3**<br>Nested urothelial carcinoma **8120/3**<br>Plasmacytoid urothelial carcinoma **8120/3**<br>Urothelial carcinoma in situ **8120/2** | Giant cell urothelial carcinoma **8031/3**<br>Lymphoepithelioma-like urothelial carcinoma **8082/3**<br>Plasmacytoid/signet ring cell/diffuse variant<br>Papillary urothelial (transitional cell) carcinoma<br>in situ **8130/2**<br>invasive **8130/3**<br>Micropapillary urothelial carcinoma **8131/3**<br>Poorly differentiated carcinoma **8020/3**<br>Sarcomatoid urothelial carcinoma **8122/3** |

## Table 3: Non-Reportable Urinary Tumors

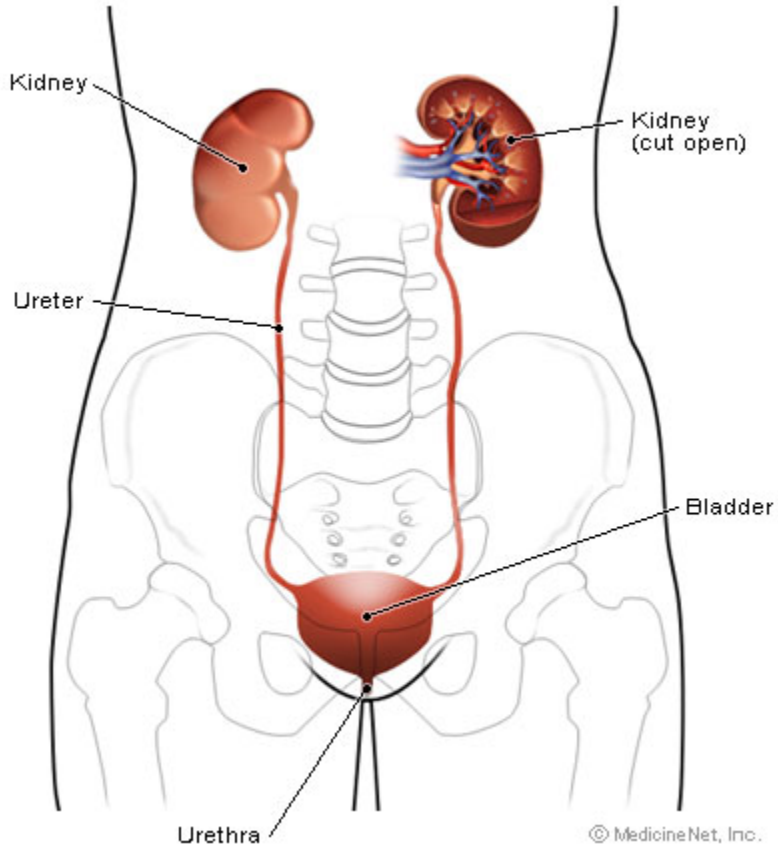
**Column 1** contains the terms and codes (if applicable) for the non-reportable histology.
**Column 2** contains **synonyms** of the histology term in column 1. Synonyms have the **same code** as the term in Column 1.

| 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



**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

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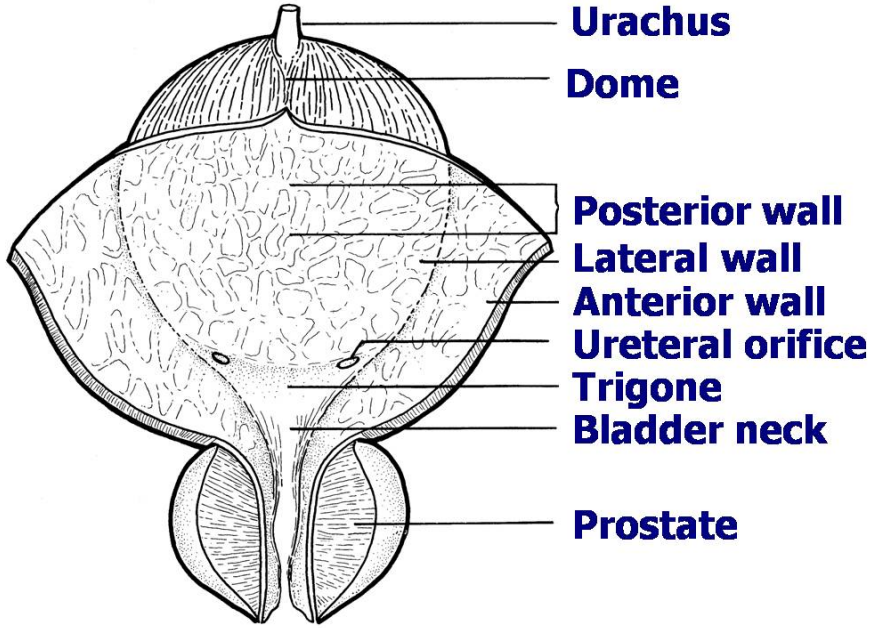
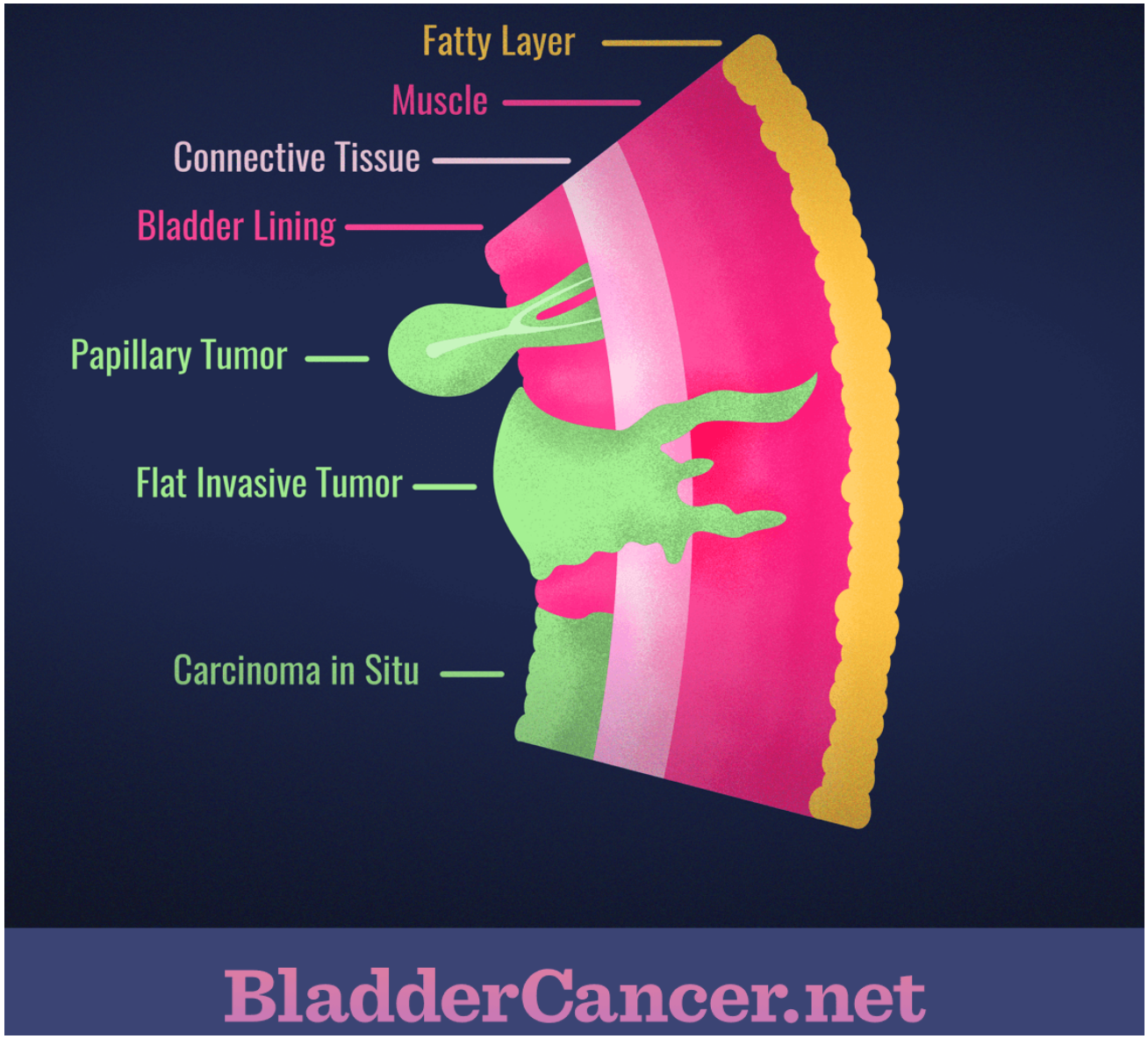



**Source: TNM Atlas, 3rd edition, 2nd revision**

**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**



**Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions**
**C659, C669, C670-C679, C680-C689**
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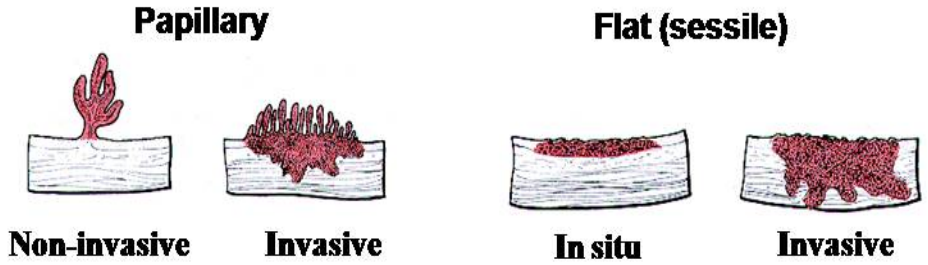
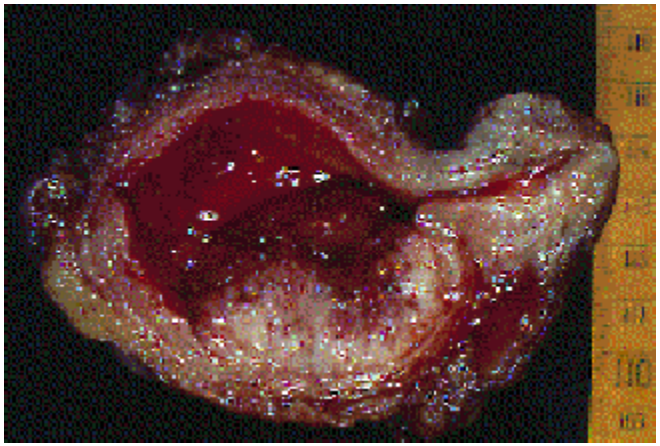
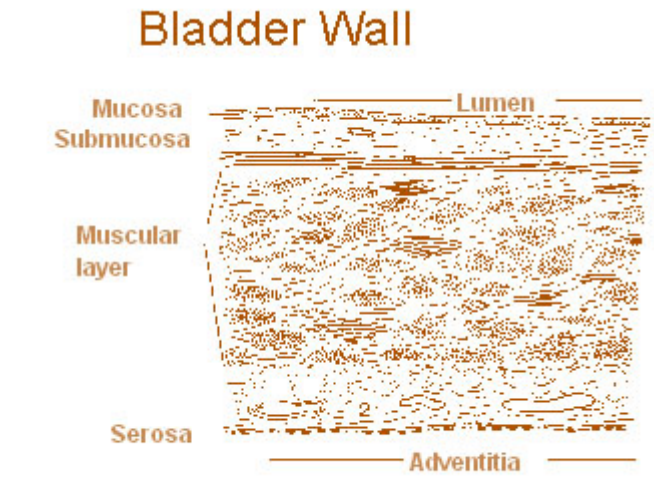


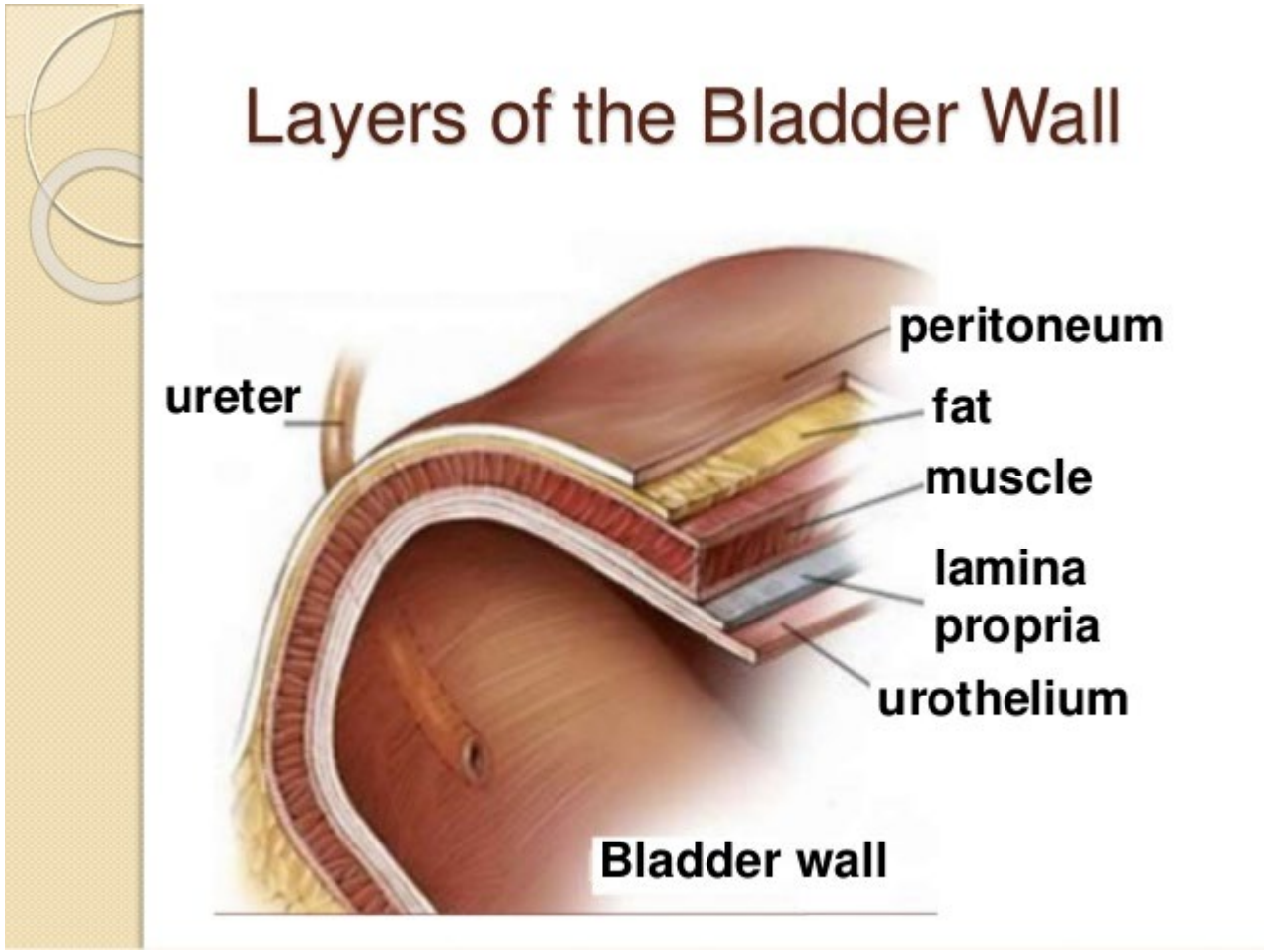
**Bladder Tumor**



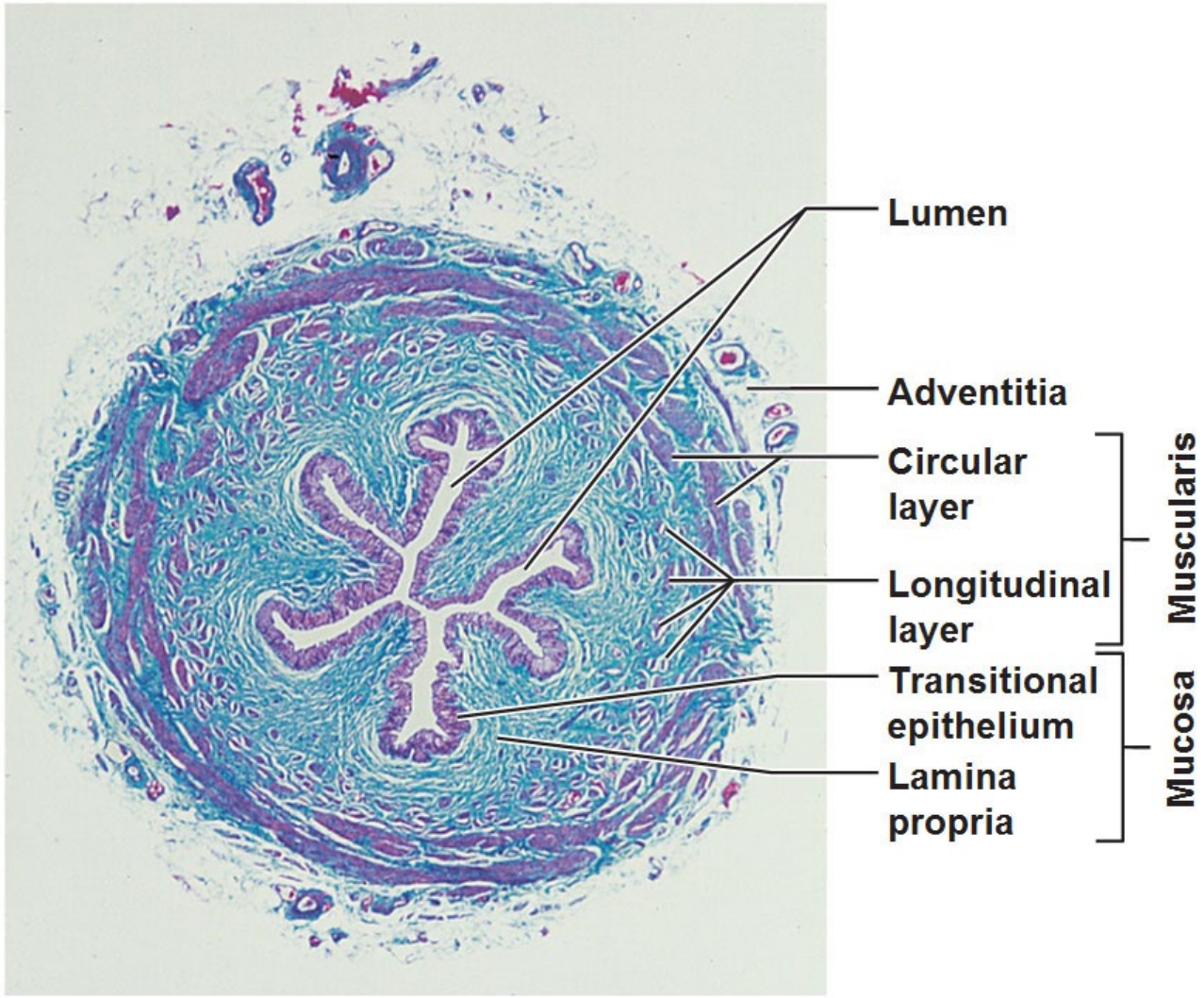
Courtesy Jean-Loup Huret reprinted from Atlas Genet Cytogenet Oncol Haematol. October 2003. van Tilborg A

A, van Rhijn BW. Bladder: Urothelial carcinomas. URL: http://atlasgeneticsoncology.org/Tumors/bladID5001.html, by permission of the Atlas.


Bladder Wall
Mucosa
Submucosa
Lumen
Muscular layer
Serosa
Adventitia
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 – M9992 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 – M9992 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 – M9992 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 – M9992 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).

**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.

***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 – M9992 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 – M9992 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 – M9992 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.
   **Exceptions:**

***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 – M9992 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

**Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules**
**C659, C669, C670-C679, C680-C689**
**(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

## 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.

**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 – M9992 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

## 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**<br>Skin of lower lip<br>Skin of upper lip | Skin of lip, NOS **C440** | **No** |
| **Eyelid**<br>Lid, NOS<br>Palpebra<br>Horizontal palpebra fissure<br>Canthus<br>Inner canthus<br>Lateral canthus<br>Lower lid<br>Medial canthus<br>Meibomian gland<br>Outer canthus<br>Pretarsal space<br>Supratarsel crease<br>Upper lid | Eyelid **C441** | **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 |
|---|---|---|
| **External ear**<br>Auricle, NOS<br>Pinna<br>Ceruminal gland<br>Concha<br>Ear, NOS<br>Ear lobule<br>Earlobe<br>External auditory canal<br>Auditory canal, NOS<br>Auricular canal, NOS<br>External auricular canal<br>Ear canal<br>External auditory meatus<br>Helix<br>Skin of auricle<br>Skin of ear, NOS<br>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**<br>Skin of:<br>Cheek<br>Chin<br>Face<br>Forehead<br>Jaw<br>Nose<br>Temple<br>Ala nasi<br>Chin, NOS<br>Columnella<br>Eyebrow<br>Brow<br>External cheek<br>External nose<br>Forehead, NOS<br>Lid-cheek junction<br>Nasaljugal groove<br>Temple, NOS | Skin of other and unspecified parts of face **C443** | **Yes** |
| **Skin of scalp and neck**<br>Skin of head, NOS<br>Skin of neck<br>Skin of scalp<br>Scalp, NOS<br>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**<br>Skin of:<br>Abdomen<br>Abdominal wall<br>Anus<br>Axilla<br>Back<br>Breast<br>Buttock<br>Chest<br>Chest wall<br>Flank<br>Groin<br>Perineum<br>Thoracic wall<br>Thorax<br>Trunk<br>Umbilicus<br>Gluteal region<br>Infraclavicular region<br>Inguinal region<br>Sacrococcygeal region<br>Scapular region<br>Perianal skin<br>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**<br>Skin of:<br>Antecubital space<br>Arm<br>Elbow<br>Finger<br>Forearm<br>Hand<br>Palm<br>Shoulder<br>Thumb<br>Upper limb<br>Wrist<br>Fingernail<br>Nail Bed<br>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**<br>Skin of:<br>Ankle<br>Calf<br>Foot<br>Heel<br>Hip<br>Knee<br>Leg<br>Lower limb<br>Popliteal space<br>Thigh<br>Toe<br>Plantar skin<br>Sole of foot<br>Toenail | Skin of lower limb and hip **C447** | **Yes** |
| **Overlapping lesion of skin** | Overlapping lesion of skin **C448**<br>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**<br><br>***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**

**Table 2: Specific Histologies, NOS, and Subtypes/Variants**

| NOS Histology Terms and Codes | Synonyms | Subtypes/Variants |
|---|---|---|
| Melanoma, NOS **8720**<br><br>***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**<br>Early/Evolving melanoma in situ** **8720/2**<br>Nevoid melanoma **8720/3**<br>Early/Evolving invasive melanoma** **8720/3** | Acral melanoma*/acral lentiginous melanoma, malignant **8744/3**<br>Amelanotic melanoma **8730/3**<br>Balloon cell melanoma **8722/3**<br>Desmoplastic melanoma/desmoplastic melanoma, amelanotic/neurotropic melanoma, malignant **8745/3***<br>Epithelioid cell melanoma **8771/3**<br>Lentigo maligna/Hutchinson melanotic freckle **8742/2**<br>Lentigo maligna melanoma/Melanoma in Hutchinson melanotic freckle **8742/3**<br>Low cumulative sun damage melanoma*/superficial spreading melanoma **8743/3**<br>Melanoma arising in a blue nevus **8780/3***<br>Malignant melanoma arising in giant congenital nevus*/malignant melanoma in giant pigmented nevus **8761/3**<br>Malignant melanoma in a precancerous melanosis **8741/3**<br>Malignant melanoma, regressing **8723/3**<br>Malignant Spitz tumor*/mixed epithelioid and spindle cell melanoma **8770/3**<br>Nodular melanoma **8721/3**<br>Spindle cell melanoma **8772/3**<br>Spindle cell melanoma, type A **8773/3**<br>Spindle cell melanoma, type B **8774/3** |

## 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<br>Nevus, NOS<br>Melanocytic nevus<br>Hairy nevus<br>Nevus spilus<br>Meyerson nevus<br>Deep penetrating nevus<br>Combined nevus<br>Genital nevus<br>Conjunctival nevus | **8720/0** |
| Balloon cell nevus | **8722/0** |
| Halo nevus<br>Regressing nevus | **8723/0** |
| Neuronevus | **8725/0** |
| Magnocellular nevus<br>Melanocytoma, NOS | **8726/0** |
| Dysplastic nevus | **8727/0** |
| Nonpigmented nevus<br>Achromic nevus | **8730/0** |
| Junctional nevus, NOS<br>Intraepidermal nevus<br>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**

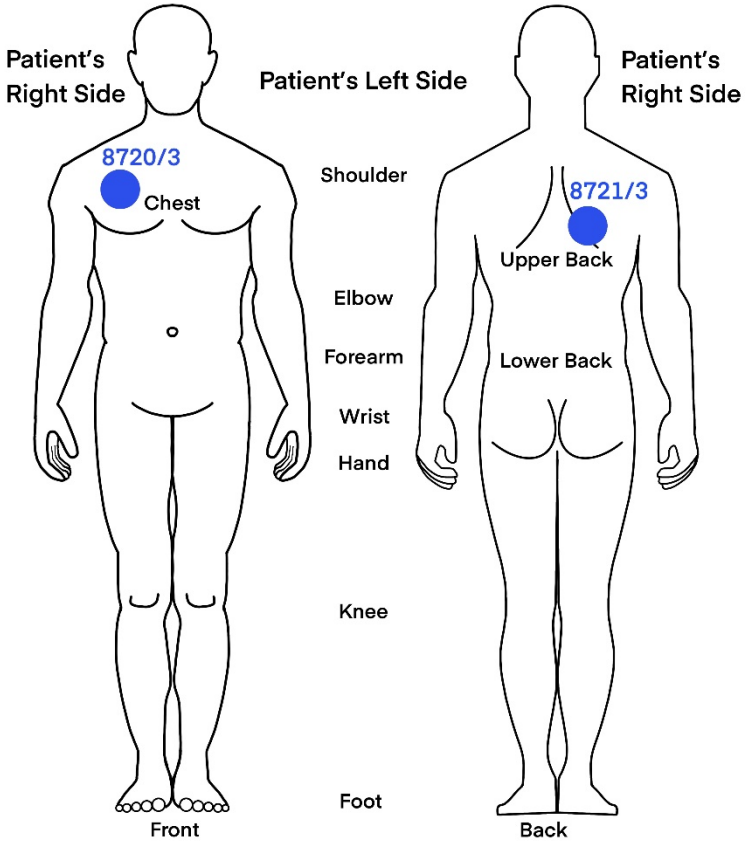
| Non-Reportable Histology Term | Non-Reportable Histology Code |
|---|---|
| Lentiginous melanocytic nevus<br>Simple lentigo<br>Lentigo simplex | **8742/0** |
| Acral nevus | **8744/0** |
| Dermal nevus<br>Intradermal nevus<br>Stromal nevus | **8750/0** |
| Compound nevus<br>Dermal and epidermal nevus | **8760/0** |
| Congenital melanocytic nevus, NOS<br>Giant pigmented nevus, NOS<br>Intermediate and giant congenital nevus | **8761/0**<br>**8761/0**<br>**8761/1** |
| Proliferative dermal lesion in congenital nevus<br>Proliferative nodule in congenital melanocytic nevus | **8762/1** |
| Epithelioid and spindle cell nevus<br>Juvenile melanoma<br>Juvenile nevus<br>Spitz nevus<br>Spitz nevus, atypical<br>Pigmented spindle cell nevus of Reed<br>Pigmented spindle cell Spitz nevus | **8770/0** |
| Epithelioid cell nevus | **8771/0** |
| Spindle cell nevus, NOS | **8772/0** |
| Blue nevus, NOS<br>Jadassohn blue nevus<br>Pigmented epithelioid melanocytoma<br>Blue nevus, epithelioid | **8780/0**<br>**8780/0**<br>**8780/1**<br>**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<br>Melanocytic neoplasm of low malignant potential<br>Melanocytic tumor of uncertain malignant potential (MELTUMP)<br>Superficial atypical melanocytic proliferation of uncertain significance (SAMPUS)<br>Primary acquired melanosis | No ICD-O code |

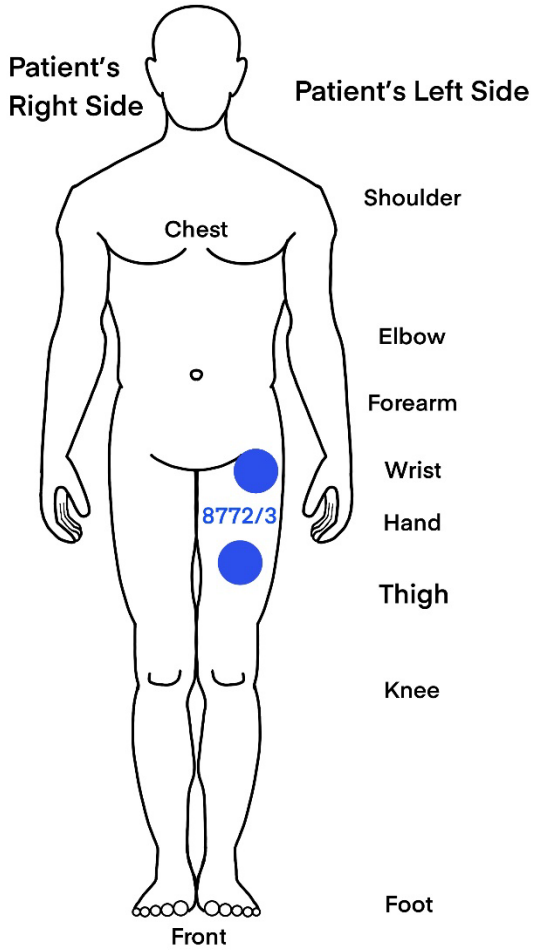
## Illustrations

### 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.

**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.

**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.

**Rule H8** When two or more melanoma subtype/variants are present in a single tumor, 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 rare.

***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.**

---

**[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.**

The following section is to be used for Other Sites cases diagnosed 2007-2022.

The 2007 General Instructions are to be used in conjunction with the Other Sites section. DO NOT USE the Solid Tumor 2018 General Instructions for these cases.

# IV.
# General Instructions and Histology Type ICD-O-3

## EQUIVALENT OR EQUAL TERMS

Adenocarcinoma, glandular carcinoma
Multicentric, multifocal
Tumor, mass, lesion, neoplasm

## DEFINITIONS

***Note*: Use these terms and definitions for all reportable cases except lymphoma and leukemia primaries (M9590-9989).**

**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.

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

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

**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)

**Most representative specimen:** The pathologic specimen from the surgical procedure that removed the most **tumor** tissue.

**Multiple primaries:** More than one reportable case.

**Overlapping tumor:** The involved sites are adjacent (next to each other) and the tumor is contiguous.

**Paired organ/site:** There are two sides, 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:

1. 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.
2. A new occurrence of cancer arising 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.

**Single primary:** One reportable case.

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

## DETERMINING MULTIPLE PRIMARIES FOR SOLID MALIGNANT TUMORS

***Note:*** The rules do not apply to hematopoietic primaries (lymphoma and leukemia) of any site or to the reportable benign or borderline intracranial or CNS tumors.

### A. General Information

1. Use these rules to determine the number of reportable primaries. Do **not** use these rules to determine case reportablility, stage, or grade.
2. The 2007 multiple primary and histology coding rules **replace all previous** multiple primary and histology coding **rules**.
3. The rules are **effective** for cases **diagnosed January 1, 2007** and after. Do not use these rules to abstract cases diagnosed prior to January 1, 2007.
4. Read the **General Instructions** and the **site-specific Equivalent Terms and Definitions** before using the multiple primary rules.
5. The multiple primary and histology coding rules are available in **three formats**: flowchart, text, and matrix. The **rules are identical**, only the formats differ. Use the rules in the format that is easiest for you to follow.
6. **Notes** and **examples** are included with some of the rules to **highlight key points** or to add **clarity** to the rules.
7. **Do not use** a physician's statement to decide whether the patient has a recurrence of a previous cancer or a new primary. Use the multiple primary rules as written **unless** a **pathologist compares** the present tumor to the "original" tumor and states that this tumor is a recurrence of cancer from the previous primary.
8. Use the Determining Multiple Primaries: Hematopoietic Primaries (Lymphoma and Leukemia) rules and table "Definitions of Single and Subsequent Primaries for Hematologic Malignancies" to determine single versus multiple primaries for lymphoma and leukemia cases.

### B. How to Use the Multiple Primary Rules

1. Use the **Multiple Primary** rules to **make a decision on the number of primary malignancies** to be abstracted for reportable solid malignant tumors.
2. Use the site-specific rules for the following primary sites:
   - Brain, malignant (intracranial and CNS)
   - Breast

- Colon
- Head and neck
- Kidney
- Lung
- Malignant melanoma of the skin
- Renal pelvis, ureter, bladder, and other urinary

3. Use the **Other Sites rules** for solid malignant tumors that occur in primary sites not covered by the site-specific rules.
4. Each module (Unknown if Single or Multiple Tumors, Single Tumor, Multiple Tumors) is an independent, complete set of coding rules. To determine which set of primary site rules to use:
   a. When there is no tumor in the primary site, only metastatic lesions are present:
      I. Use the primary site documented by a physician and use the multiple primary and histology coding rules for that primary site.
      II. If no primary site is documented, code the primary site as unknown and use the general multiple primary and histology coding rules. Use the "Unknown if Single or Multiple Tumors" module to determine multiple primaries and the "Single Tumor" module for coding histology.
   b. To choose the appropriate module (Unknown if Single or Multiple Tumors, Single Tumor, Multiple Tumors),
      I. Use the multiple primary and histology coding rules for the primary site
      II. Determine the number of tumors
         i. Do not count metastatic lesions
         ii. When the tumor is only described as multicentric or multifocal and the number of tumors is not mentioned, use the "Unknown if Single or Multiple Tumors" module
         iii. When there is a tumor or tumors with separate microscopic foci, ignore the separate microscopic foci and use the "Single Tumor" or "Multiple Tumor" modules as appropriate
         iv. When the patient has a single tumor, use the "Single Tumor" module.
         v. If there are multiple tumors, use the "Multiple Tumor" module.
      III. See the Equivalent Terms and Definitions for Head and Neck for guidance in coding the primary site
      IV. Use the primary site documented by the physician on the medical record
5. If a **single primary**, prepare **one abstract**.
6. If there are **multiple primaries**, prepare **two or more abstracts**.
7. Rules are in hierarchical order within each module (Unknown if Single or Multiple Tumors, Single Tumor, and Multiple Tumors). Use the first rule that applies and

# STOP

## 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. It affects the 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.

**Information about the 2007 Histology Coding Rules**

***Note:*** Do not use these rules to determine case reportability.

1. The 2007 multiple primary rules **replace all previous** multiple primary **rules**.
2. The rules are **effective** for cases **diagnosed January 1, 2007** and after. Do not use these rules to abstract cases diagnosed prior to January 1, 2007.
3. The histology coding rules are available in **three formats**: flowchart, text, and matrix. The **rules are identical**, only the formats differ. Use the set of rules in the format that is easiest for you to follow.
4. **Notes** and **examples** are included with some of the rules to **highlight key points** or to add **clarity** to the rules.
5. Rules are in **hierarchical** order within each section (Single Tumor and Multiple Tumors Abstracted as a Single Primary).

**How to Use the Rules**

1. Read the **General Instructions**.
2. Read the **site-specific Equivalent Terms and Definitions**.
3. Use these rules to make a decision on coding the histology for all reportable solid malignant tumors.
4. Use the multiple primary rules to determine whether the patient has a single or multiple primaries before coding the histology.
5. Code the histology for **each** primary in a **separate abstract.**
6. Use the **site-specific rules** for the following primary sites:
    - Brain, malignant (intracranial and CNS)
    - Breast
    - Colon
    - Head and neck
    - Kidney
    - Lung
    - Malignant melanoma of the skin

- Renal pelvis, ureter, bladder, and other urinary

7. Use the **Other Sites rules** for all solid malignant tumors that occur in primary sites **not included** in the site-specific rules.
8. Determine whether the patient has a single tumor or multiple tumors that will be abstracted as a single primary
   a. Do not count metastatic tumors
   b. When the tumor is described as multifocal or multicentric, use the Multiple Tumors module
   c. When there is a tumor or tumors with separate foci of tumor do not count the foci
   d. Only count the tumors that will be used to prepare that abstract. For example, when there are two tumors that will be abstracted as multiple primaries, you would use the Single Tumor modules to determine the histology code for each of the abstracts.
9. **Each section** (Single Tumor and Multiple Tumors Abstracted as a Single Primary) is an independent, **complete set of coding rules**. For example, if the patient has multiple tumors, that will be abstracted as a single primary start with the first rule under the heading Multiple Tumors Abstracted as a Single Primary. Do not use any of the rules under the header Single Tumor.
10. Use the first rule that applies and

# STOP

**Priority order for using Documents to Code Histology**

Medical records frequently include multiple pathology reports and references to histologic diagnosis. Use the following instructions to identify which reports best represent the histology to be coded.

1. Pathology report:
   a. From the **most representative** tumor specimen examined
   b. From the **final diagnosis**
      ***Note 1:*** Use information from **addenda** and **comments** associated with the final diagnosis to code the histology.
      ***Note 2:*** A **revised/amended diagnosis** replaces the original final diagnosis. Code the histology from the revised/amended diagnosis.
      ***Note 3:*** The new rules **limit** the information **to the final diagnosis**. The old rules allowed coding from information in the microscopic description. You will only use information from the microscopic portion of the pathology report when instructed to do so in one of the site-specific rules.
2. Cytology report.
3. When you do not have either a pathology report or cytology report:
   a. Documentation in the medical record that references pathology or cytology findings
   b. From mention of type of cancer (histology) in the medical record

## Ambiguous Terms Used to Code Histology

When any of the ambiguous terms are used to describe a more specific histology, code the more specific histology.

**Ambiguous terms that are characteristic (used to code histology)**

Apparent(ly)
Appears
Comparable with
Compatible with
Consistent with
Favor(s)
Most likely
Presumed
Probable
Suspect(ed)
Suspicious (for)
Typical (of)

***Example:*** Non-small cell carcinoma, most likely adenocarcinoma. Code adenocarcinoma.

## General Instructions Histology Coding Rules

When using rule (see note) that states "Code the histology documented by the physician when the pathology/cytology report is not available" code the histology from the document with the highest priority. Make a second pass through the histology rules to determine which histology code should be recorded. Start with the appropriate module, Single Tumor or Multiple Tumors, and continue through the rules until you reach the rule that fits the case you are coding.

***Note:*** For most sites this will be rule H1 and the first rule in the Multiple Tumors module

When using rule (see note) that states "When the only histology is from a metastatic site" make a second pass through the histology rules to determine which histology code should be recorded. Start with the appropriate module, Single Tumor or Multiple Tumors, and continue through the rules until you reach the rule that fits the case you are coding.

***Note:*** For most sites this will be rule H2 and the second rule in the Multiple Tumors module

When the patient has a previous or subsequent unknown primary site (80.9) or an ill-defined primary site, check carefully to see if this abstract or document should be consolidated into the previous abstract rather than making it a new primary.

# 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

## Introduction

**For cases diagnosed 1/1/2007 to 12/31/2022**

**IMPORTANT INFORMATION ON SITES COVERED IN THIS MODULE:**

**For cases diagnosed 1/1/2007 to 12/31/2017:**
The Other Sites Rules cover rectosigmoid, rectum and all sites not included in the site-specific rules.

**For cases diagnosed 1/1/2018 forward, the following sites are no longer included in the Other Sites Rules:**
Rectosigmoid C199
Rectum C209
Peripheral Nerves and Autonomic Nervous System C470-C479

Beginning 1/1/2018, rectosigmoid (C199) and rectum (C209) primaries are included in the 2018 Colon Solid Tumor Rules
Beginning 1/1/2018, peripheral nerves (C470-C479) are included in the Malignant CNS and Peripheral Nerves Solid Tumor Rules

## Equivalent or Equal Terms

- Acinar adenocarcinoma, adenocarcinoma (For prostate primaries only)
- Adenocarcinoma, glandular carcinoma
- And; with
  ***Note:*** "And" and "with" are used as synonyms when **describing multiple histologies** within a **single tumor**.
- 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

**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**

## Table 1: Paired Organs and Sites with Laterality

***Note:*** This table only includes anatomic sites covered by the Other Sites Rules.

| 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 9) |
| C445 | Skin of the trunk (if midline, assign code 9) |
| 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 |
| 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 |
| C754 | Carotid body |

## Table 2: Mixed and Combination Codes

**This table is used to determine mixed and combination codes ONLY**
Apply the Multiple Primary Rules FIRST. Combination codes are most often used when multiple histologies are present in a single tumor; they are rarely used for multiple tumors. Use a combination code for multiple tumors ONLY when the tumors meet the rules for a single primary.

Use this **two-page** table to select combination histology codes. Compare the terms in the diagnosis to the terms in Columns 1 and 2. If the terms match, code the case using the ICD-O-3 histology code in column 4. Use the combination codes listed in this table only when the histologies in the tumor match the histologies listed below.

| **Column 1:**<br>**Required Histology** | **Column 2:**<br>**Combined with Histology** | **Column 3:**<br>**Combination Term** | **Column 4:**<br>**Code** |
|---|---|---|---|
| Small cell carcinoma | Large cell carcinoma<br>Adenocarcinoma<br>Squamous cell carcinoma | Combined small cell carcinoma | 8045 |
| Squamous carcinoma | Basal cell carcinoma | Basosquamous carcinoma | 8094 |
| Islet cell | Exocrine | Mixed islet cell and exocrine adenocarcinoma (pancreas) | 8154 |
| Acinar | Endocrine | Mixed islet cell and exocrine adenocarcinoma (pancreas) | 8154 |
| Hepatocellular carcinoma | Cholangiocarcinoma | Combined hepatocellular carcinoma and cholangiocarcinoma | 8180 |
| Adenocarcinoma | Carcinoid | Composite carcinoid | 8244 |
| Adenocarcinoma and **two or more** of the histologies from column 2 OR<br>**two or more** of the histologies from column 2 | Papillary<br>Clear cell<br>Mucinous (colloid)<br>Signet ring<br>Acinar | Adenocarcinoma with mixed subtypes<br>Adenocarcinoma combined with other types of carcinoma | 8255 |

**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**

| **Column 1:**<br>**Required Histology** | **Column 2:**<br>**Combined with Histology** | **Column 3:**<br>**Combination Term** | **Column 4:**<br>**Code** |
|---|---|---|---|
| Gyn malignancies with two or more of the histologies in column 2<br><br>***Note***: First refer to ICD-O-3.2 and ICD-O updates to confirm if the mixed histology has a specific code. Example: papillary serous adenocarcinoma is coded 8460 per ICD-O. | Clear cell<br>Endometrioid<br>Mucinous<br>Papillary<br>Serous<br>Squamous<br>Transitional (Brenner) | Mixed cell adenocarcinoma | 8323 |
| Papillary and<br>Follicular | | Papillary carcinoma, follicular variant | 8340 |
| Medullary | Follicular | Mixed medullary-follicular carcinoma | 8346 |
| Medullary | Papillary | Mixed medullary-papillary carcinoma | 8347 |
| Squamous carcinoma and<br>Adenocarcinoma | | Adenosquamous carcinoma | 8560 |
| Any combination of histologies in Column 2 | Myxoid<br>Round cell<br>Pleomorphic | Mixed liposarcoma | 8855 |
| Embryonal rhabdomyosarcoma | Alveolar rhabdomyosarcoma | Mixed type rhabdomyosarcoma | 8902 |
| Teratoma | Embryonal carcinoma | Teratocarcinoma | 9081 |
| Teratoma and one or more of the histologies in Column 2 | Seminoma<br>Yolk sac tumor | Mixed germ cell tumor | 9085 |
| Choriocarcinoma | Teratoma<br>Seminoma<br>Embryonal | Choriocarcinoma combined with other germ cell elements | 9101 |

## Table 3: Changes to Previous SEER Site Grouping Table

Previous to 2007, tumors in sites on the same row were abstracted as a single primary.

| Code | Site Groupings |
|---|---|
| C23 | Gallbladder |
| C24 | Other and unspecified parts of the biliary tract |
| C37 | Thymus |
| C380 | Heart |
| C381-3 | Mediastinum |
| C388 | Overlapping lesion of heart, mediastinum, and pleura |
| C51 | Vulva |
| C52 | Vagina |
| C577 | Other specified female genital organs |
| C578-9 | Unspecified female genital organs |
| C569 | Ovary |
| C570 | Fallopian tube |
| C571 | Broad ligament |
| C572 | Round ligament |
| C573 | Parametrium |
| C574 | Uterine adnexa |
| C60 | Penis |
| C63 | Other and unspecified male genital organs |
| C74 | Adrenal gland |
| C75 | Other endocrine glands and related structures |

# 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 1/1/2007 to 12/31/2022**

**IMPORTANT INFORMATION ON SITES COVERED IN THESE MODULES:**

**For cases diagnosed 1/1/2007 to 12/31/2017:**
The Other Sites Rules cover rectosigmoid, rectum and all sites not included in the site-specific rules.

**For cases diagnosed 1/1/2018 forward, the following sites are no longer included in the Other Sites Rules:**
Rectosigmoid C199
Rectum C209
Peripheral Nerves and Autonomic Nervous System C470-C479 (Specifically rule M12)

Beginning 1/1/2018, rectosigmoid (C199) and rectum (C209) primaries are included in the 2018 Colon Solid Tumor Rules
Beginning 1/1/2018, peripheral nerves (C470-C479) are included in the Malignant CNS and Peripheral Nerves Solid Tumor Rules

## Unknown if Single or Multiple Tumors

***Note:*** These rules are **NOT** used for tumor(s) described as metastases.

**Rule M1** When it is not possible to determine if there is a **single** tumor **or multiple** tumors, opt for a single tumor and abstract as a single primary. *

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

*** Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.**
**This is the end of instructions for Unknown if Single or Multiple Tumors.**

## Other Sites Multiple Primary Rules
## Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

### Single Tumor

***Note 1:*** These rules are **NOT** used for tumor(s) described as metastases.
***Note 2:*** Includes combinations of in situ and invasive

**Rule M2** A **single tumor** is always a single primary. *
***Note:*** The tumor may overlap onto or extend into adjacent/contiguous site or subsite.

*** Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.**
**This is the end of instructions for Single Tumor.**

### Multiple Tumors

Multiple tumors may be a single primary or multiple primaries.
***Note 1:*** These rules are **NOT** used for tumor(s) described as metastases.
***Note 2:*** Includes combinations of in situ and invasive

**Rule M3** **Adenocarcinoma** of the **prostate** is always a single primary. *
***Note 1:*** Report only one adenocarcinoma of the prostate per patient per lifetime.
***Note 2:*** 95% of prostate malignancies are the common (acinar) adenocarcinoma histology (8140). See Equivalent Terms, Definitions and Tables for more information.
***Note 3:*** If patient has a previous acinar adenocarcinoma of the prostate in the database and is diagnosed with adenocarcinoma in 2007 it is a single primary.

**Rule M4** Retinoblastoma is always a single primary (unilateral or bilateral). *

**Rule M5** **Kaposi sarcoma** (any site or sites) is always a single primary. *

**Rule M6** **Follicular and papillary** tumors in the **thyroid** within 60 days of diagnosis are a single primary. *

## Other Sites Multiple Primary Rules
## Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

**Rule M7** **Bilateral epithelial** tumors (8000-8799) of the **ovary** within 60 days are a single primary. *

**Rule M8** Tumors on **both sides** (right and left) of a site listed in Table 1 are multiple primaries. **
***Note:*** See Table 1: Paired Organs and Sites with Laterality

**Rule M9** Adenocarcinoma in adenomatous polyposis coli (**familial polyposis**) with one or more in situ or malignant polyps is a single primary.*
***Note:*** Tumors may be present in a single or multiple segments of the **colon, rectosigmoid, rectum**.

**Rule M10** Tumors diagnosed **more than one (1) year** apart are multiple primaries. **

**Rule M11** 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. **
***Example 1*:** A tumor in the penis C**60**9 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 cervix 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 M12** 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. **
- Anus and anal canal (C21_)
- Bones, joints, and articular cartilage (C40_- C41_)
- Peripheral nerves and autonomic nervous system (C47_) (Cases diagnosed 1/1/2007 to 12/31/2017 ONLY)
- Connective subcutaneous and other soft tissues (C49_)
- Skin (C44_)

**Rule M13** A **frank** in situ or malignant **adenocarcinoma** and an in situ or **malignant** tumor in a **polyp** are a single primary.*

**Rule M14** **Multiple** in situ and/or **malignant polyps** are a single primary. *
***Note:*** Includes all combinations of adenomatous, tubular, villous, and tubulovillous adenomas or polyps.

## Other Sites Multiple Primary Rules
## Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

**Rule M15** An **invasive** tumor **following** an **in situ** tumor more than 60 days after diagnosis 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.

**Rule M16** Abstract as a single primary* when one tumor is:

- Cancer/malignant neoplasm, NOS (8000) and another is a specific histology or
- Carcinoma, NOS (8010) and another is a specific carcinoma or
- Squamous cell carcinoma, NOS (8070) and another is specific squamous cell carcinoma or
- Adenocarcinoma, NOS (8140) and another is a specific adenocarcinoma or
- Melanoma, NOS (8720) and another is a specific melanoma
- Sarcoma, NOS (8800) and another is a specific sarcoma

**Rule M17** Tumors with ICD-O-3 **histology** codes that are **different** at the first (**x**xxx), second (x**x**xx) or third (xx**x**x) number are multiple primaries. **

**Rule M18** Tumors that **do not meet any** of the above **criteria** are a single primary. *

***Note:*** When an invasive tumor follows an in situ tumor within 60 days, abstract as a single primary.

*** Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.**
**** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.**
**This is the end of instructions for Multiple Tumors.**

# 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 1/1/2007 to 12/31/2022**

**IMPORTANT INFORMATION ON SITES COVERED IN THESE MODULES:**

**For cases diagnosed 1/1/2007 to 12/31/2017:**
The Other Sites Rules cover rectosigmoid, rectum and all sites not included in the site-specific rules.

**For cases diagnosed 1/1/2018 forward, the following sites are no longer included in the Other Sites Rules:**
Rectosigmoid C199
Rectum C209
Peripheral Nerves and Autonomic Nervous System C470-C479

Beginning 1/1/2018, rectosigmoid (C199) and rectum (C209) primaries are included in the Colon Solid Tumor Rules
Beginning 1/1/2018, peripheral nerves (C470-C479) are included in the Malignant CNS and Peripheral Nerves Solid Tumor Rules

**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 the histology
- Documentation in the medical record that refers to 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 (cancer/malignant neoplasm, NOS) or 8010 (carcinoma, 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:*** 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.

**Rule H3** Code **8210** (adenocarcinoma in **adenomatous polyp**), **8261** (adenocarcinoma in **villous adenoma**), or **8263** (adenocarcinoma in **tubulovillous adenoma**) when:

- The final diagnosis is adenocarcinoma in a polyp or
- The final diagnosis is adenocarcinoma **and** a residual polyp or polyp architecture is recorded in other parts of the pathology report or
- The final diagnosis is adenocarcinoma **and** there is reference to a residual or pre-existing polyp or
- The final diagnosis is mucinous/colloid or signet ring cell adenocarcinoma in a polyp or
- There is documentation that the patient had a polypectomy

***Note:*** It is important to know that the adenocarcinoma originated in a polyp.

**Rule H4** **Code the** most **specific** histologic **term** when the diagnosis is:

- Carcinoma in situ, NOS (8010) and a specific in situ carcinoma or
- Squamous cell carcinoma in situ, NOS (8070) and a specific in situ squamous cell carcinoma or
- Adenocarcinoma in situ, NOS (8140) and a specific in situ adenocarcinoma or
- Melanoma in situ, NOS (8720) and a specific in situ melanoma
- Sarcoma, NOS (8800) and a specific sarcoma

***Note:*** The specific histology may be identified as type, subtype, predominantly, with features of, major, with differentiation, architecture or pattern. The terms architecture and pattern are subtypes only for in situ cancer.

**Rule H5** Code the appropriate combination/mixed code (Table 2) when there are **multiple specific histologies** or when there is a non-specific histology **with multiple specific histologies**

***Note:*** The specific histology may be identified as type, subtype, predominantly, with features of, major, with ___ differentiation, architecture or pattern. The terms architecture and pattern are subtypes only for in situ cancer.

**Rule H6** Code the histology with the **numerically higher** ICD-O-3 code.

**This is the end of instructions for a Single Tumor: In Situ Carcinoma Only.**
**Code the histology according to the rule that fits the case.**

## Single Tumor: Invasive And In Situ
## (Both invasive and in situ components)

**Rule H7** Code the single invasive histology**. Ignore the in situ** terms.

***Note:*** This is a change from the previous histology coding rules and is different from ICD-O-3 rules. This change was made in collaboration with the ICD-O-3 editors. The consensus was that coding the invasive component of the tumor better explains the likely disease course and survival category.

**This is the end of instructions for a Single Tumor: Invasive and In Situ Carcinoma.**
**Code the histology according to the rule that fits the case.**

## Single Tumor: Invasive Only
## (All parts are invasive)

**Rule H8** Code the histology documented by the physician when there is **no pathology/cytology specimen** or the **pathology/cytology** report is **not available**.

***Note 1:*** Priority for using documents to code the histology

- Documentation in the medical record that refers to pathologic or cytologic findings
- Physician's reference to type of cancer (histology) in the medical record
- CT, PET, or MRI scans

***Note 2:*** Code the specific histology when documented.

***Note 3:*** Code the histology to 8000 (cancer/malignant neoplasm, NOS) or 8010 (carcinoma, NOS) as stated by the physician when nothing more specific is documented.

**Rule H9** Code the histology from a metastatic site when there is **no pathology/cytology specimen from the primary site**.

***Note:*** Code the behavior /3.

**Rule H10** Code 8140 (adenocarcinoma, NOS) for prostate primaries when the diagnosis is acinar (adeno)carcinoma.

## Other Sites Histology Rules
## Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast, Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

**Rule H11** 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:*** If this is a papillary carcinoma of the thyroid, go to Rule H14.

**Rule H12** Code **8210** (adenocarcinoma in **adenomatous polyp**), **8261** (adenocarcinoma in **villous adenoma**), or **8263** (adenocarcinoma in **tubulovillous adenoma**) when any of the following are true:

- The final diagnosis is adenocarcinoma in a polyp
- The final diagnosis is adenocarcinoma and a residual polyp or polyp architecture is recorded in other parts of the pathology report
- The final diagnosis is adenocarcinoma and there is reference to a residual or pre-existing polyp
- The final diagnosis is adenocarcinoma mucinous/colloid or signet ring cell adenocarcinoma in a polyp
- There is documentation that the patient had a polypectomy

***Note:*** It is important to know that the adenocarcinoma originated in a polyp.

**Rule H13** **Code the** most **specific** histologic **term.** Examples include:

- Cancer/malignant neoplasm, NOS (8000) **and** a more specific histology or
- Carcinoma, NOS (8010) **and** a more specific carcinoma or
- Squamous cell carcinoma, NOS (8070) **and** a more specific squamous cell carcinoma or
- Adenocarcinoma, NOS (8140) **and** a more specific adenocarcinoma or
- Melanoma, NOS (8720) **and** a more specific melanoma or
- Sarcoma, NOS (8800) **and** a more specific sarcoma

***Note:*** The specific histology may be identified as type, subtype, predominantly, with features of, major, or with ___ differentiation. The terms architecture and pattern are subtypes only for in situ cancer.

***Example 1:*** Adenocarcinoma, predominantly mucinous. Code mucinous adenocarcinoma 8480.

***Example 2:*** Non-small cell carcinoma, papillary squamous cell. Code papillary squamous cell carcinoma 8052.

**Rule H14** Code papillary carcinoma of the thyroid to papillary adenocarcinoma, NOS (8260).

**Rule H15** Code follicular and papillary carcinoma of the thyroid to papillary carcinoma, follicular variant (8340).

**Rule H16** Code the appropriate combination/mixed code (Table 2) when there are **multiple specific histologies** or when there is a non-specific histology **with multiple specific histologies**

***Note:*** The specific histologies may be identified as a type, subtype, predominantly, with features of, major, or with ____ differentiation.

***Example 1 (multiple specific histologies):*** Mucinous and papillary adenocarcinoma. Code 8255 (adenocarcinoma with mixed subtypes)

***Example 2 (multiple specific histologies):*** Combined small cell and squamous cell carcinoma. Code 8045 (combined small cell carcinoma)

***Example 3 (non-specific with multiple specific histologies):*** Adenocarcinoma with papillary and clear cell features. Code 8255 (adenocarcinoma with mixed subtypes)

**Rule H17** Code the histology with the **numerically higher** ICD-O-3 code.

**This is the end of instructions for a Single Tumor: Invasive Carcinoma Only.**
**Code the histology according to the rule that fits the case.**

## Multiple Tumors Abstracted as a Single Primary

**Rule H18** Code the histology documented by the physician when there is **no** pathology/cytology specimen or the **pathology/cytology** report is **not available**.

***Note 1:*** Priority for using documents to code the histology

- From reports or notes in the medical record that document or reference pathologic or cytologic findings
- From clinician reference to type of cancer (histology) in the medical record
- CT, PET or MRI scans

***Note 2:*** Code the specific histology when documented.

***Note 3:*** Code the histology to 8000 (cancer/malignant neoplasm, NOS) or 8010 (carcinoma, NOS) as stated by the physician when nothing more specific is documented.

**Rule H19** Code the histology from a metastatic site when there is **no pathology/cytology specimen from the primary site**.

***Note:*** Code the behavior /3.

**Rule H20** Code 8140 (adenocarcinoma, NOS) for prostate primaries when the diagnosis is acinar (adeno)carcinoma.

**Rule H21** Code 8077/2 (Squamous intraepithelial neoplasia, grade III) for in situ squamous intraepithelial **neoplasia grade III** in sites such as the **vulva** (VIN III) **vagina** (VAIN III), or **anus** (AIN III).

***Note 1:*** VIN, VAIN, and AIN are squamous cell carcinomas. Code 8077 cannot be used for glandular intraepithelial neoplasia such as prostatic intraepithelial neoplasia (PIN) or pancreatic intraepithelial neoplasia (PAIN).

***Note 2:*** This code may be used for reportable-by-agreement cases.

**Rule H22** Code 8148/2 (Glandular intraepithelial neoplasia grade III) for in situ glandular **intraepithelial neoplasia grade III** in sites such as the **pancreas** (PAIN III).

***Note:*** This code may be used for reportable-by-agreement cases such as intraepithelial neoplasia of the **prostate** (PIN III).

**Rule H23** 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.

**Rule H24** 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 H25** Code **8210** (adenocarcinoma in **adenomatous polyp**), **8261** (adenocarcinoma in **villous adenoma**), or **8263** (adenocarcinoma in **tubulovillous adenoma**) when any of the following are true:

- The final diagnosis is adenocarcinoma in a polyp
- The final diagnosis is adenocarcinoma **and** a residual polyp or polyp architecture is recorded in other parts of the pathology report
- The final diagnosis is adenocarcinoma **and** there is reference to a residual or pre-existing polyp
- The final diagnosis is mucinous/colloid or signet ring cell adenocarcinoma in a polyp
- There is documentation that the patient had a polypectomy

***Note:*** It is important to know that the adenocarcinoma originated in a polyp.

**Rule H26** Code papillary carcinoma of the thyroid to papillary adenocarcinoma, NOS (8260).

**Rule H27** Code **follicular** and **papillary** carcinoma of the **thyroid** to papillary carcinoma, follicular variant (8340).

**Rule H28** Code the single invasive histology for **combinations** of **invasive** and **in situ.** Ignore the in situ terms.

***Note:*** This is a change from the previous histology coding rules and is different from ICD-O-3 rules. This change was made in collaboration with the ICD-O-3 editors. The consensus was that coding the invasive component of the tumor better explains the likely disease course and survival category.

**Rule H29** **Code** the most **specific** histologic **term.** Examples include:

- Cancer/malignant neoplasm, NOS (8000) **and** a more specific histology
- Carcinoma, NOS (8010) **and** a more specific carcinoma
- Squamous cell carcinoma, NOS (8070) **and** a more specific squamous cell carcinoma
- Adenocarcinoma, NOS (8140) **and** a more specific adenocarcinoma
- Melanoma, NOS (8720) **and** a more specific melanoma
- Sarcoma, NOS (8800) **and** a more specific sarcoma

***Note:*** The specific histology may be identified as type, subtype, predominantly, with features of, major, or with ___ differentiation. The terms architecture and pattern are subtypes only for in situ cancer.

***Example 1:*** Adenocarcinoma, predominantly mucinous. Code mucinous adenocarcinoma 8480.

***Example 2:*** Non-small cell carcinoma, papillary squamous cell. Code papillary squamous cell carcinoma 8052.

**Rule H30** Code the appropriate combination/mixed code (Table 2) when there are **multiple specific histologies** or when there is a non-specific histology **with multiple specific histologies.**

***Note:*** The specific histologies may be identified as a type, subtype, predominantly, with features of, major, or with ____ differentiation.

***Example 1 (multiple specific histologies):*** Gyn malignancy with mucinous, serous and papillary adenocarcinoma. Code 8323 (mixed cell adenocarcinoma)

***Example 2 (multiple specific histologies):*** Combined small cell and squamous cell carcinoma. Code 8045 (combined small cell carcinoma)

***Example 3 (non-specific with multiple specific histologies):*** Adenocarcinoma with papillary and clear cell features. Code 8255 (adenocarcinoma with mixed subtypes)

**Rule H31** Code the histology with the **numerically higher** ICD-O-3 code.

**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.**