Solid Tumor Rules

Effective with Cases Diagnosed 1/1/2018 and Forward

Updated July 2019

Editors: Lois Dickie, CTR, NCI SEER
Carol Hahn Johnson, BS, CTR (Retired), Consultant
Suzanne Adams, BS, CTR (IMS, Inc.)
Serban Negoita, MD, PhD, CTR, NCI SEER

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.

Peggy Adamo, NCI SEER
Theresa Anderson, Canada
Mari Carlos, USC/SEER
Louanne Currence, Missouri
Frances Ross, Kentucky/SEER
Raymundo Elido, Hawaii/SEER
Jim Hofferkamp, NAACCR
Meichin Hsieh, Louisiana/SEER
Carol Kruchko, CBTRUS
Bobbi Matt, Iowa/SEER
Pamela Moats, West Virginia
Patrick Nicolin, Detroit/SEER
Cathy Phillips, Connecticut/SEER

Elizabeth Ramirez, New Mexico/SEER
Monika Rivera, New York
Jennifer Ruhl, NCI SEER
Nancy Santos, Connecticut/SEER
Kacey Wigren, Utah/SEER
Carolyn Callaghan, Seattle/SEER
Shawky Matta, California/SEER
Mignon Dryden, California/SEER
Linda O’Brien, Alaska/SEER
Mary Brandt, California/SEER
Sarah Manson, CDC
Lynda Douglas, CDC
Angela Martin, NAACCR
The 2007 Multiple Primary and Histology (MPH) Coding Rules have been revised and are now referred to as 2018 Solid Tumor Rules. The 2007 Multiple Primary and Histology Coding Rules were developed to promote consistent and standardized coding by cancer registrars and the 2018 Solid Tumor Rules continue to provide coding instructions to ensure accurate data collection. It is important to note that eight site specific coding modules have been updated for 2018. These site groups are: Benign Brain, Malignant CNS, Breast, Colon, Lung, Head & Neck, Kidney, and Urinary. The remaining two site specific coding modules have not been updated for 2018. These site groups are: Cutaneous Melanoma and Other sites.

The primary reference for both the 2007 MPH rules and 2018 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 2018 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 2018 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 have been revised for 2018. The 2018 General Instructions apply ONLY 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

The 2007 Multiple Primary & Histology rules and the 2007 General Instructions are to be used for cases diagnosed 1/1/2007 to 12/31/2020 for the following site groups:
- Cutaneous melanoma
  - Cutaneous melanoma site rules will be revised for 2021 implementation to incorporate information from the new WHO 4th Ed Tumors of Skin scheduled to be released in 2018
- Other Sites
  - Primary sites excluded are:
    - Rectosigmoid and rectum which are included in 2018 Colon rules
    - Peripheral nerves which are included in 2018 Malignant Brain rules
  - Other Sites rules will be revised for 2021 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)
- Majority; major; predominantly; greater than 50%
- Multicentric; multifocal
- Simultaneous; synchronous; at the same time; prior to first course treatment
- Topography; site code
- Tumor; mass; tumor mass; lesion; neoplasm; nodule
  o 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
  o These terms are used ONLY to determine multiple primaries
  o 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.
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 1/1/2007 through 12/31/2020:
   - Cutaneous Melanoma (not updated for 2018)
   - Other Sites (not updated for 2018) for solid tumors which occur in primary sites not covered by the site-specific rules.

5. 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 (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

6. 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 #6B.
      iv. If multiple primaries, follow the priority order in #6B 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

7. The Solid Tumor Rules are available in text format.
8. Notes and examples are included with some of the rules to highlight key points or to add clarity to the rules.
9. 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).
General Instructions: Solid Tumor Rules 2018
(Excludes lymphoma and leukemia M9590 – M9992)

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.

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 evidence** 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”.

### Histologic Type ICD-O-3

<table>
<thead>
<tr>
<th>Item Length: 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>NAACCR Item #: 522</td>
</tr>
<tr>
<td>NAACCR Name: Histologic Type ICD-O-3</td>
</tr>
</tbody>
</table>

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 North American Association of Central Registries (NAACCR) has released Guidelines for ICD-O-3 Histology Code and Behavior Update effective for cases diagnosed 1/1/2018 forward. The update includes:

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

Since a release date for either ICD-O-3.2 or ICD-O-5 is unknown, the Solid Tumor Editors recommend coding histology using:

- The 2018 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

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

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

- Case is accessioned (added to your database) based on a single histology described by ambiguous terminology and no other histology information is available/documentled

**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.

<table>
<thead>
<tr>
<th>Ambiguous Terminology</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Apparently</td>
<td>Most likely</td>
</tr>
<tr>
<td>Appears</td>
<td>Presumed</td>
</tr>
<tr>
<td>Comparable with</td>
<td>Probable</td>
</tr>
<tr>
<td>Compatible with</td>
<td>Suspect(ed)</td>
</tr>
<tr>
<td>Consistent with</td>
<td>Suspicious (for)</td>
</tr>
<tr>
<td>Favor(s)</td>
<td>Typical (of)</td>
</tr>
<tr>
<td>Malignant appearing</td>
<td></td>
</tr>
</tbody>
</table>

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.
General Instructions

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.

**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.

**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”.

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.
      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
- Majority; major; predominantly; greater than 50%
- Simultaneous; synchronous; existing at the same time; concurrent; prior to first course treatment
Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

- Topography; site code
- Tumor; mass; tumor mass; lesion; neoplasm
  - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician’s statement** that the term is **malignant/cancer**
  - These terms are used **ONLY** to determine multiple primaries
  - **Do not** use these terms for casefinding or determining reportability
- Type; subtype; variant

**Terms that are NOT Equivalent or Equal**

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

- **Phenotype** is not equivalent to **subtype/type/variant**

**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**.

**Table begins on next page**

Jump to **Multiple Primary Rules**
Jump to **Histology Rules**
# Breast Equivalent Terms and Definitions

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

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

C500-C506, C508-C509

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

<table>
<thead>
<tr>
<th>Terms and Descriptive Language</th>
<th>Site Term and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inferior inner&lt;br&gt; Inferior medial&lt;br&gt; Lower inner quadrant (LIQ) &lt;br&gt; Lower medial</td>
<td>Lower inner quadrant of breast C503</td>
</tr>
<tr>
<td>Superior lateral&lt;br&gt; Superior outer&lt;br&gt; Upper lateral&lt;br&gt; Upper outer quadrant (UOQ)</td>
<td>Upper outer quadrant of breast C504</td>
</tr>
<tr>
<td>Inferior lateral&lt;br&gt; Inferior outer&lt;br&gt; Lower lateral&lt;br&gt; Lower outer quadrant (LOQ)</td>
<td>Lower outer quadrant of breast C505</td>
</tr>
<tr>
<td>Axillary tail of breast&lt;br&gt; Tail of breast NOS&lt;br&gt; Tail of Spence</td>
<td>Axillary tail of breast C506</td>
</tr>
</tbody>
</table>
| 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 C508<br> 
*Note: This is a single tumor which overlaps quadrants/subsite.*
### Terms and Descriptive Language

<table>
<thead>
<tr>
<th>Terms and Descriptive Language</th>
<th>Site Term and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>¾ or more of breast involved with tumor</td>
<td>Breast NOS <strong>C509</strong></td>
</tr>
<tr>
<td>Diffuse (tumor size 998)</td>
<td></td>
</tr>
<tr>
<td>Entire breast</td>
<td></td>
</tr>
<tr>
<td>Inflammatory without palpable mass</td>
<td></td>
</tr>
<tr>
<td>Multiple tumors in different subsites (quadrants) within the same breast</td>
<td></td>
</tr>
</tbody>
</table>

**Note:** Used for:
- Non-contiguous *multiple* tumors in *different quadrants/subsites* of same breast OR
- *Unknown/unable to identify* in which *quadrant/subsite* the tumor is *located* (Example: Outpatient biopsy with no quadrant identified. Patient lost to follow-up.)
- Inflammatory carcinoma; diffuse tumor
Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – 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.

Table begins on next page

Jump to [Multiple Primary Rules](#)
Jump to [Histology Rules](#)
### Breast Equivalent Terms and Definitions

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

#### Required Histology Terms

<table>
<thead>
<tr>
<th>DCIS/duct carcinoma/carcinoma NST 8500</th>
<th>AND</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lobular carcinoma 8520</td>
<td></td>
</tr>
</tbody>
</table>

**Note 1:** Both histologies, duct and lobular, **must have** the *same behavior* code.

**Note 2:** 8522 is used when:
- Duct **AND** lobular carcinoma are present in a *single tumor* OR
- Duct is present in at least one tumor and lobular is present in at least one tumor in the *same breast* OR
- One tumor is mixed duct and lobular; the other tumor in the *same breast* is either duct or lobular OR
- All tumors in the *same breast* are mixed duct and lobular

**Example:** One tumor with invasive duct CA in LOQ RT breast; second tumor with invasive lobular in UOQ RT breast

**Note 3:** **Do not** use 8522 when the diagnosis is carcinoma NST/duct carcinoma with lobular differentiation. See [Histology Rules](#) for instructions on coding differentiation.

<table>
<thead>
<tr>
<th>DCIS/duct carcinoma/carcinoma NST OR any ONE subtype/variant of carcinoma NST</th>
<th>AND</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Any</strong> histology in Table 3 with <strong>exception</strong> of</td>
<td></td>
</tr>
<tr>
<td>• Lobular carcinoma 8520 and pleomorphic lobular carcinoma in situ 8519/2*</td>
<td></td>
</tr>
<tr>
<td>• Paget disease 8540</td>
<td></td>
</tr>
</tbody>
</table>

**Note 1:** Both histologies **must have** the *same behavior* code.

**Note 2:** See Table 3 for carcinoma NST/duct carcinoma subtypes/variants.

**Note 3:** **Do not** use combination code for duct with lobular differentiation. This is a synonym for carcinoma NST.

### Histology Combination Term and Code

<table>
<thead>
<tr>
<th>Invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma 8522/3</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Note 1:</strong> CAP uses the term Invasive carcinoma with ductal and lobular features (“mixed type carcinoma”)</td>
</tr>
<tr>
<td><strong>Note 2:</strong> Carcinoma NST includes all subtypes/variants of carcinoma NST.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>DCIS and in situ lobular carcinoma 8522/2</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Note:</strong> The lobular carcinoma includes pleomorphic lobular carcinoma in situ 8519/2.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Invasive carcinoma NST/duct mixed with other types of invasive carcinoma 8523/3</th>
</tr>
</thead>
<tbody>
<tr>
<td>DCIS mixed with other in situ carcinoma 8500/2</td>
</tr>
<tr>
<td><strong>Note:</strong> Prior to 2018, DCIS and other in situ was coded 8523/2.</td>
</tr>
</tbody>
</table>

---

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Rules](#)
<table>
<thead>
<tr>
<th>Required Histology Terms</th>
<th>Histology Combination Term and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Lobular carcinoma</strong></td>
<td>Infiltrating lobular mixed with other types of carcinoma 8524/3</td>
</tr>
</tbody>
</table>
| **Any** history in **Table 3 with exception** of  
  - Duct carcinoma/carcinoma NST/DCIS (and subtypes/variants) 8500  
  - Paget disease, in situ and invasive  |
| **Lobular carcinoma and Paget**  | Paget disease (invasive or behavior not specified) and DCIS/intraductal carcinoma 8543/3  
  Paget disease (specified as in situ) and DCIS/intraductal carcinoma 8543/2  |
| **Paget disease**        | Paget disease and infiltrating duct carcinoma 8541/3 |
| **Underlying** DCIS      | Paget disease and infiltrating duct carcinoma 8541/3 |
| **Paget disease**        | Paget disease and infiltrating duct carcinoma 8541/3 |
| **Underlying infiltrating duct** carcinoma/carcinoma NST and all subtypes/variants of infiltrating duct/carcinoma NST (must be a /3)  |
| **Any** two invasive carcinoma NST subtypes/variants (percentage not stated) abstracted as a single primary  
  **Note 1:** The diagnosis may be two subtypes/variants and the pathologist may mention the presence of duct/carcinoma NST. Ignore the mention of carcinoma NST.  
  **Note 2:** See **Table 3** for subtypes/variants of carcinoma NST/duct carcinoma.  | Adenocarcinoma with mixed subtypes 8255/3  |
Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – 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 using the Solid Tumor Rules, rhabdomyosarcoma and its subtypes/variants are treated the same as all NOS and subtypes/variants.

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

<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Acinic cell carcinoma 8550          | Acinar adenocarcinoma  
Acinar carcinoma               |         |
| Adenoid cystic carcinoma (ACC) 8200| ACC  
Adenocystic basal cell carcinoma  
Carcinoma adenoides cysticum  
Cylindromatous carcinoma |         |
| Adenomyoepithelioma with carcinoma 8983 | AME  
Malignant AME |         |
| Apocrine carcinoma 8401            |         | |
| **Note:** This is a diagnosis that is EXACTLY apocrine carcinoma, not a carcinoma NST with apocrine features, differentiation, or type. |
| Carcinoma NST 8500                 | Carcinoma of no special type (ductal/NST)  
Carcinoma/carcinoma NST with choriocarcinomatous features  
Carcinoma/carcinoma NST with cribriform features  
Carcinoma/carcinoma NST with melanotic features  
Carcinoma/carcinoma NST with signet ring cell differentiation  
DCIS 8500/2  
Duct/ductal carcinoma  
Duct/ductal carcinoma in situ 8500/2  
Duct/ductal carcinoma NOS | Carcinoma with osteoclastic-like stromal giant cells 8035  
Pleomorphic carcinoma 8201/3  
Pleomorphic carcinoma 8022/3 |

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Rules](#)
Breast Equivalent Terms and Definitions  
C500-C506, C508-C509  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Duct/ductal carcinoma NST (no special type) | Duct/ductal carcinoma with apocrine features  
Duct/ductal carcinoma with apocrine metaplasia  
Duct/ductal carcinoma with lobular features  
Duct/ductal carcinoma with micropapillary features  
Duct/ductal carcinoma with mucin production  
Duct/ductal carcinoma with squamous metaplasia  
Infiltrating ductal carcinoma **8500/3**  
Invasive carcinoma with micropapillary features **8500/3**  
Invasive carcinoma not otherwise specified (ductal/NOS) **8500/3**  
Invasive carcinoma NST with metaplastic features **8500/3**  
Invasive carcinoma NST/duct with medullary features **8500/3**  
Invasive carcinoma, with signet-ring cell features **8500/3**  
Invasive carcinoma of no special type (NST) **8500/3**  
Invasive carcinoma with clear cell (glycogen rich) features **8500/3** |
## Breast Equivalent Terms and Definitions

**C500-C506, C508-C509**

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

<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Invasive carcinoma, NST 8500/3</td>
<td>Invasive carcinoma, type cannot be determined 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma 8500/3</td>
<td>Invasive mammary carcinoma associated with encysted papillary carcinoma 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma NST</td>
<td>Invasive mammary carcinoma NST with lobular features 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma NST</td>
<td>Invasive mammary carcinoma NST with medullary features 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma NST</td>
<td>Invasive mammary carcinoma NST with mucinous features 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma NST</td>
<td>Invasive mammary carcinoma NST with tubulo-lobular variant 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma with apocrine features 8500/3</td>
<td>Invasive mammary carcinoma with cribriform features 8500/3</td>
<td></td>
</tr>
<tr>
<td>Invasive mammary carcinoma with tubular features 8500/3</td>
<td>Mammary carcinoma in situ 8500/2</td>
<td></td>
</tr>
<tr>
<td>Mammary carcinoma/cancer</td>
<td>Non-invasive mammary carcinoma 8500/2</td>
<td></td>
</tr>
</tbody>
</table>

<p>| Glycogen-rich clear cell carcinoma 8315 | Glycogen-rich carcinoma | Clear cell carcinoma 8310 |
| Inflammatory carcinoma 8530           |                      |                               |
| Lipid-rich carcinoma 8314             | Lipid-secreting carcinoma |</p>
<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Lobular carcinoma 8520</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lobular carcinoma in situ 8520/2</td>
<td></td>
<td>Pleomorphic lobular carcinoma in situ 8519/2*</td>
</tr>
<tr>
<td>Lobular carcinoma with cribriform features</td>
<td></td>
<td>Note: 8519/2 is a new code for in situ /2 tumors only.</td>
</tr>
<tr>
<td>Mixed lobular carcinoma (lobular carcinoma NOS and one or more variants of lobular carcinoma)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Invasive pleomorphic lobular carcinoma 8520/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Solid lobular carcinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tubulolobular carcinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Medullary carcinoma 8510</strong></td>
<td></td>
<td>Atypical medullary carcinoma (AMC) 8513</td>
</tr>
<tr>
<td>MC</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table continues on next page
Breast Equivalent Terms and Definitions  
C500-C506, C508-C509  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Metaplastic carcinoma NOS or of no special type (NST) 8575 | Invasive mammary carcinoma with matrix production  
Metaplastic carcinoma, mixed epithelial and mesenchymal type  
Metaplastic carcinoma with mesenchymal differentiation  
Metaplastic carcinoma with squamous features  
Metaplastic carcinoma with other types of mesenchymal differentiation  
Mixed metaplastic carcinoma | Carcinosarcoma 8980/3  
Fibromatosis-like metaplastic carcinoma 8572  
Low grade adenosquamous carcinoma 8570  
Metaplastic carcinoma spindle-cell type/spindle cell carcinoma 8032  
Metaplastic carcinoma with chondroid differentiation/with osseous differentiation 8571  
Myoepithelial carcinoma 8982  
Sarcomatoid carcinoma 8033  
Squamous cell carcinoma 8070 |

**Note:** 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.

| Mucinous carcinoma 8480 | Colloid carcinoma  
Mucinous adenocarcinoma  
Mucoid carcinoma |  |

**Note 1:** This is a diagnosis that is EXACTLY “mucinous carcinoma,” “mucinous duct carcinoma,” “mucinous DCIS” OR “greater than 90% mucinous.” See Histology Rules.

**Note 2:** Mucinous duct carcinoma is listed on the CAP protocol. It is not recognized by WHO or IARC. Mucinous carcinoma is not a subtype/variant of Carcinoma NST/duct carcinoma.

| Mucoepidermoid carcinoma 8430 |  |

<p>| Oncocytic carcinoma 8290 |  |</p>
<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Paget disease of the nipple with no underlying tumor 8540/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Papillary carcinoma 8503</td>
<td>Intraductal papillary carcinoma 8503/2*</td>
<td>Encapsulated papillary carcinoma 8504</td>
</tr>
<tr>
<td></td>
<td>Intraductal papillary carcinoma with DCIS 8503/2*</td>
<td>non-infiltrating/intracystic 8504/2</td>
</tr>
<tr>
<td></td>
<td>Invasive papillary carcinoma 8503/3</td>
<td>with invasion 8504/3</td>
</tr>
<tr>
<td></td>
<td>Papillary carcinoma non-invasive 8503/2*</td>
<td>Micropapillary carcinoma 8507*</td>
</tr>
<tr>
<td></td>
<td>Papillary ductal carcinoma in situ 8503/2*</td>
<td>Solid papillar carcinoma in situ 8509/2*</td>
</tr>
<tr>
<td></td>
<td></td>
<td>with invasion 8509/3</td>
</tr>
<tr>
<td>Periductal stromal tumor, low grade 9020/3</td>
<td>Phyllodes tumor, malignant</td>
<td></td>
</tr>
<tr>
<td>Polymorphous carcinoma 8525</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sarcoma NOS 8800/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Note: Rhabdomyosarcoma 8900/3 is also a NOS with the following subtypes/variants:</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Alveolar type rhabdomyosarcoma 8920/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Embryonal type rhabdomyosarcoma 8910/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pleomorphic rhabdomyosarcoma 8901/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Angiosarcoma 9120/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hemangiosarcoma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lymphangiosarcoma 9170/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malignant hemangioendothelioma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Liposarcoma 8850/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Leiomyosarcoma 8890/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Osteosarcoma 9180/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Rhabdomyosarcoma 8900/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Alveolar type 8920/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Embryonal type 8910/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pleomorphic 8901/3</td>
</tr>
</tbody>
</table>
Breast Equivalent Terms and Definitions  
C500-C506, C508-C509  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Specific and NOS/NST Terms and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sebaceous carcinoma 8410</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Secretory carcinoma 8502</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Signet ring carcinoma 8490</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| Small cell carcinoma 8041          | Carcinoid tumor of breast  
Endocrine carcinoma  
Neuroendocrine carcinoma, poorly  
differentiated | Carcinoma with  
neuroendocrine  
differentiation/Invasive  
mammary carcinoma with  
neuroendocrine features 8574/3  
Neuroendocrine tumor, well-  
differentiated 8246/3 |
| Tubular carcinoma 8211             |          |                  |

*New codes approved by IARC/WHO Committee for ICD-O
Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Illustrations

Netter illustration used with permission of Elsevier Inc. All rights reserved
Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Mammary Gland

Atlas of Human Anatomy -- Frank H. Netter
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:
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\(^1\) 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

\(^1\) 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\(^1\) when the diagnosis is inflammatory carcinoma in:
- Multiple quadrants of same breast **OR**
- Bilateral breasts

**Rule M3** Abstract a single primary\(^1\) 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

\(^1\) Prepare one abstract. Use the [histology rules](#) to assign the appropriate histology code.

Multiple Tumors

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

**Rule M4** Abstract multiple primaries\(^4\) 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 (C\(x\)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 C50\(x\) and a colon tumor C18\(x\) 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 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 when there is inflammatory carcinoma in:
- Multiple quadrants of same breast OR
- Bilateral breasts

Rule M7  Abstract multiple primaries 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 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 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** when multiple tumors 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.
**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** when a **ductal** carcinoma occurs **after a combination code** in the same breast. See the following list:
- **DCIS** following a diagnosis of:
  - DCIS + lobular carcinoma in situ 8522/2 OR
  - DCIS + in situ Paget 8543/2 OR
  - DCIS + Invasive Paget 8543/3 OR
  - DCIS mixed with other in situ 8523/2 (code used for cases diagnosed prior to 1/1/2018)
- **Invasive carcinoma NST/duct** following a diagnosis of:
  - Invasive duct + invasive lobular 8522/3 OR
  - Invasive duct + invasive Paget 8541/3 OR
  - Invasive duct + other invasive carcinoma 8523/3
Rule M12  Abstract multiple primaries 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 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)

Rule M14  Abstract multiple primaries 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 (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\(^1\) (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\(^2\) 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\(^1\) 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.

\(^1\) 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.
\(^2\) Prepare two or more abstracts. Use the histology rules to assign the appropriate histology code to each case abstracted.
Breast Histology Coding 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.
Breast Histology Coding Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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)
      
      **Note 2:** The majority may be indicated by terms such as “greater than 50%”, “major”, “majority” and “predominantly”.
      
      **Note 3:** The following terms **do not** describe the majority of tumor.

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

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

*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**

<table>
<thead>
<tr>
<th>Apparently</th>
<th>Most likely</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appears</td>
<td>Presumed</td>
</tr>
<tr>
<td>Comparable with</td>
<td>Probable</td>
</tr>
<tr>
<td>Compatible with</td>
<td>Suspect(ed)</td>
</tr>
<tr>
<td>Consistent with</td>
<td>Suspicious (for)</td>
</tr>
<tr>
<td>Favor(s)</td>
<td>Typical (of)</td>
</tr>
<tr>
<td>Malignant appearing</td>
<td></td>
</tr>
</tbody>
</table>
**Breast Histology Coding 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.

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 the histology using Table 2 when there are multiple in situ histologies (2 or more) within a single tumor.  
- Lobular and any histology other than DCIS 8524/2  
- Two or more histologies other than lobular and DCIS 8255/2  
*Note:* This rule does not include DCIS.  See previous rules.  

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

Code the histology according to the rule that fits the case  

Single Tumor: Invasive and In Situ Components  

Rule H7  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
Breast Histology Coding Rules  
C500-C506, C508-C509 
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Single Tumor: Invasive Only

Rule H8  
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 H9  
Code the underlying tumor when there is a diagnosis of inflammatory carcinoma.  
**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 H10  
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 1:** When a tumor has both mucinous carcinoma and a different histology, and mucinous is less than or equal to 90% of the tumor (or the percentage is not documented), code the other histology.

---

1 American College of Pathologists: Protocol for the Examination of Specimens From Patients With Invasive Carcinoma of the Breast: “Inflammatory carcinoma requires the presence of clinical findings of erythema and edema involving at least one-third or more of the skin of the breast”
Breast Histology Coding Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H11  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 H12  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 H13  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 H14  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 H15** 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 H16** Code the histology that comprises the majority (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 majority may be indicated by terms such as “greater than 50%”, “major”, “majority” and “predominantly”.

*Note 3:* The rules are hierarchical, so the tumors are NOT a NOS/NST and subtype/variant.

*Note 4:* 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.
Breast Histology Coding Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H17  Code a combination code when there are two histologies (two components) within a single tumor and the majority histology is unknown/not documented.

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

Note 2: The rules are hierarchical, so the tumors are NOT a NOS/NST and a single subtype/variant.

Note 3: The diagnosis may be two subtypes/variants and the pathologist may mention the presence of duct/carcinoma NST. Ignore the mention of carcinoma NST.

Note 4: Do not use a combination code when the second histology is described as features or differentiation unless it is part of the preferred term.

Note 5: The histologies may be identified as:
- Mixed histologies
- Combination histologies
- Histology 1 AND histology 2
- Histology 1 WITH histology 2

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

Code the histology according to the rule that fits the case

Multiple Tumors Abstracted as a Single Primary

Note 1: DCIS is often multifocal/multicentric; use the Single Tumor: In Situ module.

Note 2: First use the multiple primary rules to ensure that the multiple tumors are to be abstracted as a single primary.

Rule H18  Code the underlying tumor when there is a diagnosis of inflammatory carcinoma:

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 H19  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 H20  Code Paget disease and DCIS 8543/2 when there is Paget disease (specified as in situ) with underlying DCIS.

Rule H21  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 H22  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 H23  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.
Breast Histology Coding Rules
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H24  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 H25  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 a Multiple Tumors Abstracted as a Single Primary

Code the histology according to the rule that fits the case

Jump to Equivalent Terms and Definitions
Jump to Multiple Primary Rules
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

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

- 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.

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

### 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; carcinoma NOS; adenocarcinoma; adenocarcinoma NOS; intestinal type adenocarcinoma 8140
- 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.
Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

- 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 of 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

### 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**.
- **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)

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 /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.

<table>
<thead>
<tr>
<th>Specific and NOS Term and Code</th>
<th>Synonyms for Specific or NOS Term</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenocarcinoma 8140</td>
<td>Adenocarcinoma, NOS</td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma/carcinoma in a polyp NOS (now coded to 8140)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma/carcinoma in adenomatous polyp (now coded to 8140)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma/carcinoma in polypoid adenoma (now coded to 8140)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma/carcinoma in serrated adenoma (now coded to 8140)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adenocarcinoma and mucinous carcinoma, mucinous documented as less than 50% of tumor OR percentage of mucinous</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Note 1:** See Histology Rules for instructions on coding adenocarcinoma subtypes/variants arising in a polyp.

**Note 2:** When the term intestinal adenocarcinoma is used to describe a colon primary, it simply means the appearance is

Adenoid cystic carcinoma 8200
Cribriform comedo-type carcinoma/
adenoarcinoma, cribriform comedo-type 8201*
Diffuse adenocarcinoma/carcinoma 8145
Linitis plastica 8142/3
Medullary adenocarcinoma/carcinoma 8510
Micropapillary carcinoma 8265*
Mucinous/colloid adenocarcinoma/carcinoma 8480
Mucoepidermoid carcinoma 8430
Serrated adenocarcinoma 8213*
### Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions

*C180-C189, C199, C209*

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

<table>
<thead>
<tr>
<th>Specific and NOS Term and Code</th>
<th>Synonyms for Specific or NOS Term</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>similar to adenocarcinoma seen in the stomach and is coded to adenocarcinoma NOS 8140</td>
<td>unknown/not documented Adenocarcinoma and signet ring cell carcinoma, percentage of signet ring cell carcinoma documented as less than 50% of tumor OR percentage of signet ring cell carcinoma unknown/not documented Adenocarcinoma/carcinoma in tubular polyp (now coded to 8140) Adenocarcinoma/carcinoma in tubulovillous polyp (now coded to 8140) Adenocarcinoma/carcinoma in villous adenoma (now coded to 8140) Adenocarcinoma in any type of polyp Adenocarcinoma, intestinal type Adenocarcinoma and cribriform carcinoma percentage of cribriform documented as less than 50% of tumor OR percentage of cribriform carcinoma unknown/not documented Adenocarcinoma with mucinous and signet ring cell features Comedocarcinoma Intestinal adenocarcinoma</td>
<td>Signet ring cell/poorly cohesive adenocarcinoma/carcinoma 8490 Superficial spreading adenocarcinoma 8143 Tubulopapillary carcinoma 8263 Undifferentiated adenocarcinoma/carcinoma 8020</td>
</tr>
<tr>
<td>Adenosquamous carcinoma 8560</td>
<td>Mixed adenocarcinoma NOS and epidermoid carcinoma Mixed adenocarcinoma NOS and squamous cell carcinoma</td>
<td><strong>Note:</strong> This code cannot be used for adenocarcinoma subtypes/variants with squamous cell/epidermoid carcinoma</td>
</tr>
</tbody>
</table>

---

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)
### Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions

C180-C189, C199, C209  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Specific and NOS Term and Code</th>
<th>Synonyms for Specific or NOS Term</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| **Combined small cell carcinoma 8045** | Small cell carcinoma mixed with  
  • Adenocarcinoma OR  
  • Neuroendocrine carcinoma OR  
  • Any other type of carcinoma/adenocarcinoma |                     |

**Gastrinoma 8153**

**Gastrointestinal stromal tumor classified as malignant 8936/3**  
Gastrointestinal stromal tumor, malignant  
GIST, malignant

**Mixed adenoneuroendocrine carcinoma 8244**  
Adenocarcinoma mixed with high-grade large cell neuroendocrine carcinoma  
Adenocarcinoma mixed with high-grade small cell neuroendocrine carcinoma  
MANEC

**Neuroendocrine carcinoma 8246**  
NEC

**Neuroendocrine tumor Grade 1 (G1) 8240**  
Carcinoid NOS  
Low-grade neuroendocrine tumor  
NET Grade 1 (G1)  
Well differentiated neuroendocrine tumor

*Note:* When the diagnosis is exactly “carcinoid” it may be a Grade 1 or Grade 2 NET. Default is coding NET Grade 1 8240.

**Sarcoma NOS 8800/3**  
Angiosarcoma/hemangiosarcoma 9120/3  
Leiomyosarcoma 8890/3

**Spindle cell carcinoma 8032**

**Squamous cell carcinoma 8070**  
Epidermoid carcinoma NOS  
Squamous cell carcinoma NOS  
Squamous cell epithelioma

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

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtype/Variant of NOS with Histology Code</th>
<th>Reason not reportable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenoma 8140/0</td>
<td>Adenoma NOS</td>
<td>Tubular adenoma 8211/0 Tubulovillous adenoma 8263/0 Villous adenoma 8261/0</td>
<td>Non-malignant</td>
</tr>
<tr>
<td><strong>Note:</strong> No malignancy in polyps</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cowden-associated polyp No code</td>
<td>Cowden disease Cowden syndrome Multiple hamartoma syndrome</td>
<td></td>
<td>Non-malignant /no code</td>
</tr>
<tr>
<td><strong>Note:</strong> No malignancy in polyps</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dysplasia, high grade 8148/2</td>
<td>High-grade dysplasia Intraepithelial neoplasia, high grade</td>
<td></td>
<td>CURRENTLY NOT REPORTABLE</td>
</tr>
<tr>
<td><strong>Note:</strong> Colorectal primaries only (C180-C189, C199 and C209)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dysplasia, low grade 8148/0*</td>
<td>Intraepithelial neoplasia, low grade</td>
<td></td>
<td>Non-malignant</td>
</tr>
<tr>
<td><strong>Note:</strong> Colorectal primaries only (C180-C189, C199 and C209)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions

**C180-C189, C199, C209**

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

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtype/Variant of NOS with Histology Code</th>
<th>Reason not reportable</th>
</tr>
</thead>
</table>
| Familial adenomatous polyposis (FAP) No code | Adenomatous polyposis coli  
Bussey-Garder polyposis  
Familial multiple polyposis  
Familial polyposis coli  
Familial polyposis of the colon and rectum  
Familial polyposis of the gastrointestinal tract  
Gardner syndrome  
Multiple adenomatosis |                                          | Reportable only when there is cancer in a polyp |
| Gangliocytic paraganglioma 8683/0 |                                                                           |                                             | Non-malignant |
| Gastrointestinal stromal tumor 8936/1 | GIST NOS  
GIST, behavior not specified |                                             | Non-malignant |
| Hyperplastic polyp No code |                                                                         |                                             | Non-malignant/no code |
| Inflammatory or pseudopoly No code |                                                                         |                                             | Reactive lesions; mimic carcinoma |
| Juvenile polyp No code | Combined juvenile polyposis/hereditary  
Hemorrhagic telangiectasis (Osler-Webec-Rendu) syndrome  
Familial juvenile polyposis  
Generalized juvenile polyposis  
Hamartomatous gastrointestinal polyposis;  
Juvenile polyposis  
Juvenile polyposis coli  
Juvenile polyposis of infancy |                                             | Non-malignant / no code |
<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtype/Variant of NOS with Histology Code</th>
<th>Reason not reportable</th>
</tr>
</thead>
<tbody>
<tr>
<td>L cell glucagon-like peptide and PP/PYY-producing NETs 8152/1*</td>
<td></td>
<td></td>
<td>Non-malignant</td>
</tr>
<tr>
<td>Leiomyoma 8890/0</td>
<td></td>
<td></td>
<td>Non-malignant</td>
</tr>
<tr>
<td>Lipoma 8850/0</td>
<td></td>
<td></td>
<td>Benign accumulation of fat cells that are circumscribed or encapsulated</td>
</tr>
<tr>
<td>Low-grade appendiceal mucinous neoplasm 8480/1</td>
<td>LAMN</td>
<td></td>
<td>Non-malignant</td>
</tr>
<tr>
<td>Note: May have low-grade, non-invasive pseudomyxoma peritonei, mucinous implants in peritoneum or beyond</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lynch syndrome No code</td>
<td></td>
<td></td>
<td>Non-malignant/no code</td>
</tr>
<tr>
<td>Mesenchymal tumors</td>
<td></td>
<td>Granular cell tumor 9580/0 Hemangioma 9120/0</td>
<td>Non-malignant</td>
</tr>
<tr>
<td>Peutz-Jeghers polyp No code</td>
<td>Intraepithelial neoplasia in Peutz-Jeghers polyp(s) Periorificial lentiginosis Peutz-Jeghers polyposis Polyps-and-spots syndrome</td>
<td></td>
<td>Non-malignant/no code</td>
</tr>
</tbody>
</table>
Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions  
C180-C189, C199, C209  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtype/Variant of NOS with Histology Code</th>
<th>Reason not reportable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pseudomyxoma peritonei (when pathologist does not designate as malignant OR implants are benign) 8480/1</td>
<td></td>
<td></td>
<td>Non-malignant. When both implants and site of origin are benign, the case is not reportable.</td>
</tr>
</tbody>
</table>
| Sessile serrated adenoma/polyp 8213/0* | Serrated polyposis  
Sporadic serrated polyps  
Traditional serrated adenoma | | Non-malignant |
| Note: No malignancy in polyps | | | |
| Tubular carcinoid, no malignancy 8245/1 | | | Non-malignant |

*These new codes were approved by the IARC/WHO Committee for ICD-O
Colonoscopy measurements which may be used to determine primary site when no site is designated
Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Polyps and de novo or “frank” adenocarcinoma in colon

Source: http://upload.wikimedia.org/wikipedia/commons/4/44/Colon_cancer.jpg
Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Large intestine; snare instrument to remove polyps

![Diagram of colon sections and biopsy catheter]

Cecum  Rectum  Sigmoid colon

Transverse colon

Ascending colon  Descending colon

Biopsy catheter
Colon Surgery: Hemicolecotomy
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

Rectum

Anus

Part of rectum to be removed during surgery

Descending colon connected to remaining colon
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 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

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\(^1\) 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

\(^1\) 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\(^1\) 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.
Rule M4  Abstract multiple primaries\footnote{1} when there are separate, non-contiguous tumors in sites with ICD-O site codes that differ at the second C\textsuperscript{X}xx and/or third C\textsuperscript{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\textsuperscript{50}x and colon C\textsuperscript{18}x

*Example 2:* Colon C\textsuperscript{18}x and rectum C\textsuperscript{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\footnote{1} 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\footnote{1} 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\footnote{1} 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 24 months after original tumor resection OR
- The subsequent tumor arises in the mucosa

*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: These rules are hierarchical. Only use this rule when previous rules do not apply.

Rule M8 Abstract a single primary when a subsequent tumor arises at the anastomotic site AND:
- The subsequent tumor occurs less than or equal to 24 months after original tumor resection OR
- The tumor arises in colon/rectal wall and/or surrounding tissue; there is no involvement of the mucosa 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.

Rule M9 Abstract multiple primaries when there are separate, non-contiguous tumors in sites with ICD-O site codes that differ at the fourth characters C18X.

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.

Rule M10 Abstract multiple primaries 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.
Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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\(^1\) 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\(^1\) (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.

Rule M13 Abstract a single primary\(^1\) (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.
Colon, Rectosigmoid, and Rectum Multiple Primary Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M14  Abstract multiple primaries\(^i\) 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.

\(^i\) 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.

\(^i\) Prepare two or more abstracts. Use the histology rules to assign the appropriate histology code to each case abstracted.
IMPORTANT NOTES

   
   Note 1: Histology changes do occur following immunotherapy, chemotherapy and radiation therapy.
   
   Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

2. Code the histology assigned by the physician. 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
Colon, Rectosigmoid, and Rectum Histology Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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/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
Colon, Rectosigmoid, and Rectum Histology Rules

C180-C189, C199, C209

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

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

**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**

<table>
<thead>
<tr>
<th>Apparently</th>
<th>Most likely</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appears</td>
<td>Presumed</td>
</tr>
<tr>
<td>Comparable with</td>
<td>Probable</td>
</tr>
<tr>
<td>Compatible with</td>
<td>Suspect(ed)</td>
</tr>
<tr>
<td>Consistent with</td>
<td>Suspicious (for)</td>
</tr>
<tr>
<td>Favor(s)</td>
<td>Typical (of)</td>
</tr>
<tr>
<td>Malignant appearing</td>
<td></td>
</tr>
</tbody>
</table>

Jump to Equivalent Terms and Definitions
Jump to Multiple Primary Rules
4. **Do not code** histology when described as:

- Architecture
- Foci; focus; focal
- Pattern

### 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
**Colon, Rectosigmoid, and Rectum Histology Rules**

C180-C189, C199, C209

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

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

- **Exactly “mucinous” adenocarcinoma** (no modifiers)
- **High-grade** pseudomyxoma peritonei
- **Invasive** pseudomyxoma peritonei
- **Malignant** pseudomyxoma peritonei
- Two histologies and mucinous is documented to be *greater than 50%* of the tumor

*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) AND

- The pseudomyxoma peritonei are **high-grade/invasive/malignant** OR
- Patient is treated for malignant pseudomyxoma peritonei

*Note 3:* The following are non-reportable:

- 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
Colon, Rectosigmoid, and Rectum Histology Rules  
C180-C189, C199, C209  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**Rule H6**  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 50% of tumor
  - Adenocarcinoma and signet ring cell carcinoma
    - Percentage of signet ring unknown/not documented
    - Signet ring cell documented as less than 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 H7**  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 H8**  Code the invasive histology when in situ and invasive histologies are present in the same tumor.
Colon, Rectosigmoid, and Rectum Histology Rules  
C180-C189, C199, C209  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H9  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 H10**  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.
Colon, Rectosigmoid, and Rectum Histology Rules
C180-C189, C199, C209
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H11  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
  o Less than or equal to 100 polyps are identified OR
  o 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 H12  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 H13  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 H14  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.

Jump to Equivalent Terms and Definitions
Jump to Multiple Primary Rules
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, 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 C410 and maxilla C411, have been added to the Head and Neck Rules.

3. External ear C442 has been added to the Head and Neck Rules. Basal cell carcinoma, squamous cell carcinoma, and all non-reportable neoplasms are excluded.

4. 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, C442, 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
- Hypopharynx; laryngopharynx
- Malignant hemangioendothelioma; angiosarcoma
- 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 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
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**

- **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
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Jump to Multiple Primary Rules
Jump to Histology Coding Rules
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

C000-C148, C300-C339, C410, C411, C442, C479

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

## Table Index

<table>
<thead>
<tr>
<th>Table Number</th>
<th>Table Title</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Table 1</strong></td>
<td>Tumors of Nasal Cavity C300 Paranasal Sinuses C310-C313, C318, C319</td>
</tr>
<tr>
<td><strong>Table 2</strong></td>
<td>Tumors of Nasopharynx C110, C111 (posterior wall of nasopharynx only), C112, C113, C117, C119</td>
</tr>
<tr>
<td><strong>Table 3</strong></td>
<td>Pyriform Sinus C129 Tumors of Hypopharynx C130-C132, C138, C139 Larynx C320-C323, C328, C329 Trachea C339 and Parapharyngeal Space C139</td>
</tr>
<tr>
<td><strong>Table 4</strong></td>
<td>Tumors of Oral Cavity and mobile tongue C020-C024, C028, C029, C030, C031, C039, C040, C041, C048, C049, C050-C052, C058, C059, C060-C062, C068, C069</td>
</tr>
<tr>
<td><strong>Table 5</strong></td>
<td>Tumors of Oropharynx C100-C104, C108 C109 Base of Tongue C019 Tonsils C090, C091, C098, C099 Adenoids/pharyngeal tonsil only C111</td>
</tr>
<tr>
<td><strong>Table 6</strong></td>
<td>Tumors of Salivary Glands C079, C080, C081, C088, C089</td>
</tr>
<tr>
<td><strong>Table 7</strong></td>
<td>Tumors of Odontogenic and Maxillofacial Bone (Mandible C410, Maxilla C411)</td>
</tr>
<tr>
<td><strong>Table 8</strong></td>
<td>Tumors of Ear C301 and External auditory canal C442</td>
</tr>
<tr>
<td><strong>Table 9</strong></td>
<td>Paraganglioma of Carotid body, Larynx, Middle Ear, Vagal nerve C479</td>
</tr>
<tr>
<td><strong>Table 10</strong></td>
<td>Paired Sites</td>
</tr>
</tbody>
</table>
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 using the Solid Tumor Rules, rhabdomyosarcoma and its subtypes/variants are treated the same as all NOS and subtypes/variants.

Table begins on next page
### Head and Neck Equivalent Terms and Definitions

**C000-C148, C300-C339, C410, C411, C442, C479**

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

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Adenocarcinoma 8140</strong></td>
<td>Adenocarcinoma non-intestinal type</td>
<td>Adenocarcinoma intestinal type (ITAC) <strong>8144</strong></td>
</tr>
<tr>
<td><strong>Note:</strong> Adenocarcinoma intestinal-type of the sinonasal tract is morphologically similar to adenocarcinomas of the intestines</td>
<td>Low-grade adenocarcinoma</td>
<td>Colloid-type adenocarcinoma <strong>8144</strong></td>
</tr>
<tr>
<td></td>
<td>Renal cell-like carcinomas</td>
<td>Colonic-type adenocarcinoma <strong>8144</strong></td>
</tr>
<tr>
<td></td>
<td>Seromucinous adenocarcinoma</td>
<td>Enteric-type adenocarcinoma <strong>8144</strong></td>
</tr>
<tr>
<td></td>
<td>TAC</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Terminal tubulous adenocarcinoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Tubulopapillary low-grade adenocarcinoma</td>
<td></td>
</tr>
<tr>
<td><strong>Lymphoepithelial carcinoma 8082</strong></td>
<td>LEC</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lymphoepithelioma-like carcinoma</td>
<td></td>
</tr>
<tr>
<td><strong>Malignant peripheral nerve sheath tumor 9540/3</strong></td>
<td>Malignant neurilemmoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Malignant schwannoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>MPNST</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Neurofibrosarcoma</td>
<td></td>
</tr>
<tr>
<td><strong>Mucoepidermoid carcinoma 8430</strong></td>
<td>Salivary gland-type mucoepidermoid carcinoma</td>
<td></td>
</tr>
<tr>
<td><strong>Mucosal melanoma 8720</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Myoepithelial carcinoma 8982</strong></td>
<td>Myoepithelioma, malignant</td>
<td></td>
</tr>
<tr>
<td><strong>Non-keratinizing squamous cell carcinoma 8072</strong></td>
<td>Cylindrical cell carcinoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>NKSCC</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Schneiderian carcinoma</td>
<td></td>
</tr>
<tr>
<td><strong>NUT carcinoma 8023</strong></td>
<td>Midline carcinoma of children and young adults with NUT rearrangement</td>
<td></td>
</tr>
<tr>
<td></td>
<td>NUT midline carcinoma</td>
<td></td>
</tr>
</tbody>
</table>
## Head and Neck Equivalent Terms and Definitions

C000-C148, C300-C339, C410, C411, C442, C479

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

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Olfactory neuroblastoma 9522/3</strong></td>
<td>Esthesioneuroblastoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Esthesioneurocytoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Esthesioneuroepithelioma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Olfactory placode tumor</td>
<td></td>
</tr>
<tr>
<td></td>
<td>ONB</td>
<td></td>
</tr>
<tr>
<td><strong>Primitive neuroectodermal tumor 9364</strong></td>
<td>Adult neuroblastoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ewings sarcoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Peripheral neuroblastoma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Peripheral neuroectodermal tumor</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Peripheral neuroepithelioma</td>
<td></td>
</tr>
<tr>
<td><strong>Sarcoma 8800/3</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 1:</strong> 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.</td>
<td>Angiosarcoma/hemangiosarcoma 9120/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Biphenotypic sinonasal sarcoma (BSNS)/low-grade sinonasal sarcoma with neural and myogenic features 9045/3*</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Epithelioid hemangioendothelioma 9133/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Fibrosarcoma/adult-type fibrosarcoma 8810/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Leiomyosarcoma 8890/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Rhabdomyosarcoma 8900/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Alveolar rhabdomyosarcoma 8920/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Embryonal rhabdomyosarcoma 8910/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pleomorphic rhabdomyosarcoma, adult type 8901/3</td>
<td></td>
</tr>
<tr>
<td><strong>Note 2:</strong> Rhabdomyosarcoma 8900/3 has subtypes/variants:</td>
<td>Spindle cell rhabdomyosarcoma 8912/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Alveolar rhabdomyosarcoma 8920/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Embryonal rhabdomyosarcoma 8910/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pleomorphic rhabdomyosarcoma, adult type 8901/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Synovial sarcoma/synovial cell sarcoma 9040/3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma 8802/3</td>
<td></td>
</tr>
</tbody>
</table>
### Specific or NOS Term and Code

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinonasal undifferentiated carcinoma 8020</td>
<td>Sinonasal carcinoma, undifferentiated SNUC</td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> This is an undifferentiated carcinoma of the Sinonasal tract.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Squamous cell carcinoma 8070</td>
<td>Keratinizing squamous cell carcinoma (KSCC) 8071 Sarcomatoid squamous cell carcinoma/spindle cell squamous cell carcinoma (SC-SCC) 8074</td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> Sinonasal squamous cell tumors account for about 3% of head and neck malignancies.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Teratocarcinosarcoma 9081</td>
<td>Blastoma Malignant teratoma Teratocarcinoma Teratoid carcinosarcoma</td>
<td></td>
</tr>
</tbody>
</table>

*These new codes were approved by the IARC/WHO Committee for ICD-O*
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, C442, C479

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

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenoid cystic carcinoma 8200</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chordoma 9370</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nasopharyngeal papillary adenocarcinoma 8260</td>
<td>Thyroid-like low-grade nasopharyngeal; papillary adenocarcinoma</td>
<td></td>
</tr>
<tr>
<td>Squamous cell carcinoma NOS 8070</td>
<td>Lymphoepithelial carcinoma&lt;br&gt;Undifferentiated carcinoma&lt;br&gt;Undifferentiated carcinoma with lymphoid stroma</td>
<td>Basaloid squamous cell carcinoma 8083&lt;br&gt;Keratinizing squamous cell carcinoma 8071&lt;br&gt;Non-keratinizing squamous cell carcinoma 8072</td>
</tr>
</tbody>
</table>
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.

**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>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenoid cystic carcinoma 8200</td>
<td>ACC (rare)</td>
<td></td>
</tr>
<tr>
<td>Chondrosarcoma 9220</td>
<td>Chondrosarcoma grade 2/3 Chondrosarcoma NOS</td>
<td></td>
</tr>
<tr>
<td>Liposarcoma 8850</td>
<td>Atypical lipomatous tumor Well-differentiated liposarcoma</td>
<td></td>
</tr>
<tr>
<td>Squamous cell carcinoma (SCC) 8070</td>
<td>Epidermoid carcinoma Squamous cell carcinoma NOS</td>
<td>Adenosquamous carcinoma (ASC) 8560 Basaloid squamous cell carcinoma (BSCC) 8083 Lymphoepithelial carcinoma (LEC)/lymphoepithelioma-like carcinoma 8082 Papillary squamous cell carcinoma (PSCC) 8052 Spindle cell squamous cell carcinoma (SC-SCC) 8074 Verrucous squamous cell carcinoma (VC) 8051</td>
</tr>
<tr>
<td>Well-differentiated neuroendocrine carcinoma 8240</td>
<td>Carcinoid Neuroendocrine carcinoma grade 1</td>
<td>Large cell neuroendocrine carcinoma/LCNEC 8013 Neuroendocrine carcinoma grade 2/moderately-differentiated neuroendocrine carcinoma/atypical carcinoid 8249 Small cell neuroendocrine carcinoma/small cell carcinoma/SmCC 8041</td>
</tr>
</tbody>
</table>
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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
- C024 Lingual tonsil
- 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
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kaposi sarcoma 9140</td>
<td>Kaposi disease</td>
<td></td>
</tr>
<tr>
<td>Mucoepidermoid carcinoma 8430</td>
<td>Mucoepidermoid tumor</td>
<td></td>
</tr>
<tr>
<td>Myofibroblastic sarcoma 8825</td>
<td>Myofibrosarcoma</td>
<td></td>
</tr>
<tr>
<td>Oral mucosal melanoma 8720</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Squamous cell carcinoma 8070</td>
<td>Squamous carcinoma</td>
<td>Acantholytic squamous cell carcinoma</td>
</tr>
<tr>
<td></td>
<td>Squamous cell carcinoma NOS</td>
<td>8075</td>
</tr>
</tbody>
</table>

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

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.

Jump to [Multiple Primary Rules](#)
Jump to [Histology Coding Rules](#)
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:

**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
- 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.

Table begins on next page
<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenoid cystic carcinoma 8200</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Polymorphous adenocarcinoma 8525</td>
<td>Cribiform adenocarcinoma, Polymorphous low-grade adenocarcinoma, Terminal duct carcinoma</td>
<td>Keratinizing squamous cell carcinoma 8071, Non-keratinizing squamous cell carcinoma 8072, Squamous cell carcinoma HPV-negative 8086*, Squamous cell carcinoma HPV-positive 8085*</td>
</tr>
<tr>
<td>Squamous cell carcinoma 8070</td>
<td></td>
<td>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.</td>
</tr>
</tbody>
</table>

* These new codes were approved by the IARC/WHO Committee for ICD-O
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
<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acinic cell carcinoma 8550</td>
<td>ACC</td>
<td>Basal cell adenocarcinoma 8147</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Basal cell adenocarcinoma-ex-monomorphic adenoma 8147</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malignant dermal analogue tumor 8147</td>
</tr>
<tr>
<td>Adenocarcinoma 8140</td>
<td>Adenocarcinoma NOS</td>
<td>Carcinoma ex-pleomorphic adenoma 8941</td>
</tr>
<tr>
<td></td>
<td>Unclassified adenocarcinoma</td>
<td>Clear cell carcinoma (CCC)/hyalinizing clear cell carcinoma 8310</td>
</tr>
<tr>
<td></td>
<td>Salivary gland adenocarcinoma NOS</td>
<td>Cribriform adenocarcinoma 8201</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intestinal-type adenocarcinoma 8144</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Large cell carcinoma NOS/large cell undifferentiated carcinoma 8012</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lobular carcinoma 8520</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Mucinous cystadenocarcinoma 8470</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Mucoepidermoid carcinoma (MEC)/malignant mucoepidermoid tumor 8430</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Papillary cystadenocarcinoma 8450</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Polymorphous adenocarcinoma (PAC) 8525</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Polymorphous low-grade adenocarcinoma 8525</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Terminal duct carcinoma 8525</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Salivary duct carcinoma 8500</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cribriform cystadenocarcinoma low-grade 8500/2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Ductal carcinoma/adenocarcinoma 8500</td>
</tr>
<tr>
<td></td>
<td></td>
<td>High-grade ductal carcinoma 8500</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intraductal carcinoma 8500/2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intraductal carcinoma low-grade 8500/2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Undifferentiated carcinoma 8020</td>
</tr>
</tbody>
</table>
**Head and Neck Equivalent Terms and Definitions**

C000-C148, C300-C339, C410, C411, C442, C479

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

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

* These new codes were approved by the IARC/WHO Committee for ICD-O
Table 7: Tumors of Odontogenic and Maxillofacial Bone (Mandible, Maxilla)

Table 7 lists the more common histologies for the following head and neck subsites:
- **C410** Bones of skull and face and associated joints; maxilla
- **C411** Mandible; jaw bone NOS; lower jaw bone; temporomandibular joint

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

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

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

**Column 2** contains *synonyms* for the specific or NOS term. Synonyms have the *same* histology code as the specific or NOS term.

**Column 3** contains *subtypes/variants* of the NOS histology. Subtypes/variants do not have the *same* histology code as the NOS.

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

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

*Table begins on next page*
### Specific or NOS Term and Code

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Ameloblastic carcinoma-primary type 9270/3             | AC  
Ameloblastic carcinoma  
Ameloblastic carcinoma, dedifferentiated  
Ameloblastic carcinoma, secondary type  
Primary intraosseous carcinoma NOS (PIOC)  
Sclerosing odontogenic carcinoma (SOC) | Metastasizing ameloblastoma 9310/3  
**Note:** This is an ameloblastoma which has a benign appearance but metastasizes |

<table>
<thead>
<tr>
<th>Clear cell odontogenic carcinoma 9341*</th>
<th>CCOC</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Note:</strong> Clear cell odontogenic tumors were classified as benign prior to the 2005 edition of WHO Pathology &amp; Genetics Head and Neck Tumors</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

| Ghost cell odontogenic carcinoma 9302*                | Aggressive epithelial ghost cell odontogenic tumor  
Calcifying ghost cell odontogenic carcinoma  
Carcinoma arising in calcifying odontogenic cyst  
Malignant calcifying ghost cell odontogenic tumor  
Malignant calcifying odontogenic cyst  
Malignant epithelial odontogenic ghost cell tumor |                                                                                                      |
### Odontogenic carcinosarcoma 8980/3
- Ameloblastic carcinosarcoma
- Malignant odontogenic mixed tumor
- Mixed odontogenic carcinoma

**Subtypes/Variants**
- Odontogenic sarcoma/ameloblastic fibrosarcoma 9330/3

### Sarcoma NOS 8800/3

**Note 1:** Osteosarcoma 9180/3 has subtypes/variants:
- Chondroblastic osteosarcoma 9181/3
- Intraosseous well-differentiated osteosarcoma/low-grade central osteosarcoma 9187/3
- Parosteal osteosarcoma 9192/3
- Periosteal osteosarcoma 9193/3

**Note 2:** Chondrosarcoma grade 2/3 9220/3 has a subtype/variant:
- Mesenchymal chondrosarcoma 9240/3

**Subtypes/Variants**
- Chondrosarcoma grade 2/3 9220/3
- Mesenchymal chondrosarcoma 9240/3
- Osteosarcoma/osteogenic sarcoma 9180/3
- Chondroblastic osteosarcoma 9181/3
- Intraosseous well-differentiated osteosarcoma/low-grade central osteosarcoma 9187/3
- Parosteal osteosarcoma 9192/3
- Periosteal osteosarcoma 9193/3

* These new codes were approved by the IARC/WHO Committee for ICD-O.
Table 8 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
- **C442** External ear; external auditory canal
  
  **Note:** External ear/skin excludes basal cell carcinoma, squamous cell carcinoma, and all non-reportable neoplasms.

**Note 1:** Use the Malignant Melanoma Rules for a melanoma in skin of ear.

**Note 2:** See the SEER Manual and/or COC Manual for reportability of skin tumors. Use Head and Neck Rules for reportable skin primaries of external ear C442 only.

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.

<table>
<thead>
<tr>
<th>Specific or NOS Term and Code</th>
<th>Synonyms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ceruminous adenocarcinoma 8420</td>
<td>Ceruminal adenocarcinoma</td>
</tr>
<tr>
<td><strong>Note:</strong> The primary site is ceruminous gland C442.</td>
<td></td>
</tr>
</tbody>
</table>
| Endolymphatic sac tumor 8140 | Adenocarcinoma  
Heftner tumor  
Low-grade papillary adenocarcinoma of endolymphatic sac origin |
| **Note:** The endolymphatic sac is located within the inner ear C301. |
| Squamous cell carcinoma of the middle ear 8070 | SCC  
Squamous carcinoma  
Squamous cell carcinoma NOS |
| **Note:** This neoplasm arises in the squamous epithelium within the middle ear C301. |
Table 9: Paraganglioma of Carotid Body, Larynx, Middle Ear, Vagal Nerve

Table 9 lists interim codes for paragangliomas. ICD-O-3 lists paraganglioma as 8690/0 and 8690/1. New codes and malignant behavior were proposed by the IARC/WHO Committee for ICD-O, but the new codes have not been implemented for use in 2018.

Only report these neoplasms when the pathology/tissue specifies malignant behavior /3. Change the behavior using ICD-O-3 Rule F Matrix Concept.

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
<table>
<thead>
<tr>
<th>Specific Term and Code</th>
<th>Synonyms for Specific Histology</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Carotid body paraganglioma 8690</strong></td>
<td>Carotid body tumor</td>
</tr>
<tr>
<td></td>
<td>Chemodectoma, carotid</td>
</tr>
<tr>
<td></td>
<td>Non-chromaffin paraganglioma, carotid</td>
</tr>
<tr>
<td>Note 1: This neoplasm is only reportable when</td>
<td></td>
</tr>
<tr>
<td>documented as malignant/invasive /3 behavior.</td>
<td></td>
</tr>
<tr>
<td>Note 2: Cases diagnosed as malignant in 2018</td>
<td></td>
</tr>
<tr>
<td>should be reported as 8690/3. The proposed new</td>
<td></td>
</tr>
<tr>
<td>code, 8692/3, cannot be used because it has not</td>
<td></td>
</tr>
<tr>
<td>been implemented.</td>
<td></td>
</tr>
<tr>
<td><strong>Laryngeal paraganglioma 8690</strong></td>
<td>Chemodectoma, laryngeal</td>
</tr>
<tr>
<td></td>
<td>Non-chromaffin paraganglioma, laryngeal</td>
</tr>
<tr>
<td>Note 1: This neoplasm is only reportable when</td>
<td></td>
</tr>
<tr>
<td>documented as malignant/invasive /3 behavior.</td>
<td></td>
</tr>
<tr>
<td>Note 2: Cases diagnosed as malignant in 2018</td>
<td></td>
</tr>
<tr>
<td>should be reported as 8690/3. The proposed new</td>
<td></td>
</tr>
<tr>
<td>code, 8693/3, cannot be used because it has not</td>
<td></td>
</tr>
<tr>
<td>been implemented.</td>
<td></td>
</tr>
<tr>
<td>Note 3: Vagal paraganglioma has the same</td>
<td></td>
</tr>
<tr>
<td>proposed histology code as laryngeal paragangli</td>
<td></td>
</tr>
<tr>
<td>oma. Laryngeal and vagal are in separate rows</td>
<td></td>
</tr>
<tr>
<td>to emphasize the primary site.</td>
<td></td>
</tr>
<tr>
<td><strong>Middle ear paraganglioma 8690</strong></td>
<td>Glomus jugulare tumor of middle ear</td>
</tr>
<tr>
<td></td>
<td>Glomus tympanicum</td>
</tr>
<tr>
<td></td>
<td>Jugulotympanic chemodectoma</td>
</tr>
<tr>
<td>Note 1: This neoplasm is only reportable when</td>
<td></td>
</tr>
<tr>
<td>documented as malignant/invasive /3 behavior.</td>
<td></td>
</tr>
<tr>
<td>Note 2: Cases diagnosed as malignant in 2018</td>
<td></td>
</tr>
<tr>
<td>should be reported as 8690/3. The proposed new</td>
<td></td>
</tr>
<tr>
<td>code, 8693/3, cannot be used because it has not</td>
<td></td>
</tr>
<tr>
<td>been implemented.</td>
<td></td>
</tr>
<tr>
<td><strong>Vagal paraganglioma 8690</strong></td>
<td>Glomus jugulare tumor of vagal trunk</td>
</tr>
<tr>
<td></td>
<td>Chemodectoma of vagal trunk</td>
</tr>
<tr>
<td></td>
<td>Non-chromaffin paraganglioma of vagal trunk</td>
</tr>
<tr>
<td>Note 1: This neoplasm is only reportable when</td>
<td></td>
</tr>
<tr>
<td>documented as malignant/invasive /3 behavior.</td>
<td></td>
</tr>
<tr>
<td>Note 2: Cases diagnosed as malignant in 2018</td>
<td></td>
</tr>
<tr>
<td>should be reported as 8690/3. The proposed new</td>
<td></td>
</tr>
<tr>
<td>code, 8693/3, cannot be used because it has not</td>
<td></td>
</tr>
<tr>
<td>been implemented.</td>
<td></td>
</tr>
<tr>
<td>Note 3: Vagal paraganglioma has the same</td>
<td></td>
</tr>
<tr>
<td>proposed histology code as laryngeal paragangli</td>
<td></td>
</tr>
<tr>
<td>oma. Laryngeal and vagal are in separate rows</td>
<td></td>
</tr>
<tr>
<td>to emphasize the primary site.</td>
<td></td>
</tr>
</tbody>
</table>
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.

<table>
<thead>
<tr>
<th>Paired Sites</th>
<th>Site Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frontal sinus</td>
<td>C312</td>
</tr>
<tr>
<td>Maxillary sinus</td>
<td>C310</td>
</tr>
<tr>
<td>Middle ear</td>
<td>C301</td>
</tr>
<tr>
<td>Nasal cavity (excluding nasal cartilage, nasal septum)</td>
<td>C300</td>
</tr>
<tr>
<td>Tonsil</td>
<td>C09</td>
</tr>
<tr>
<td>Parotid gland</td>
<td>C079</td>
</tr>
<tr>
<td>Skin of External Ear</td>
<td>C442</td>
</tr>
<tr>
<td>Sublingual gland</td>
<td>C081</td>
</tr>
<tr>
<td>Submandibular gland</td>
<td>C080</td>
</tr>
</tbody>
</table>
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Illustrations

A.D.A.M illustration used with licensed permission. All rights reserved
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Head and Neck Equivalent Terms and Definitions
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, C442, 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\(^1\) 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
      o Outpatient biopsy with no follow-up information available
      o 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.

\(^1\) 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, C442, 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\(^i\) when there are separate/non-contiguous tumors in any two of the following sites:
- Glottis C320 AND/OR supraglottis C321 AND/OR subglottis C322 AND/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
Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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\text{x}. Use this rule ONLY for the primary sites listed.

Rule M4 Abstract multiple primaries when separate/non-contiguous tumors are present in sites with ICD-O site codes that differ at the second C\text{X}xx, and/or third characters \text{C}x\text{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 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 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.
**Rule M7**  
Abstract **multiple primaries** 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** 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** (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:* 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 M10**  
Abstract a **single primary** (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.
Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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\(^\text{ii}\) 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\(^1\) 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)

Rule M13 Abstract a single primary\(^1\) 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.

---

\(^1\) 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.

\(^\text{ii}\) Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted

Jump to Equivalent Terms and Definitions
Jump to Histology Coding Rules
Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES
   \textit{Note 1:} Histology changes do occur following immunotherapy, chemotherapy and radiation therapy.
   \textit{Note 2:} Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

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 \textbf{most specific histology} from either \textit{resection} or \textit{biopsy}.
\textit{Note 1:} The term “most specific” usually refers to a subtype/variant.
\textit{Note 2:} The histology rules instruct to code the invasive histology when there are in situ and invasive components in a single tumor.
\textit{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. \textbf{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
   \textit{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.
   \textit{Note 2:} The pathologist’s diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.
   \textit{Note 3:} The CAP protocol is a checklist which:
      \begin{itemize}
         \item Provides guidelines for collecting the essential data elements for complete reporting of malignant tumors and optimal patient care.
         \item Allows physicians to check multiple histologies
      \end{itemize}

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, C442, 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/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/documentedy

   **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.**
Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, C442, C479
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

List of Ambiguous Terminology

<table>
<thead>
<tr>
<th>Apparently</th>
<th>Most likely</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appears</td>
<td>Presumed</td>
</tr>
<tr>
<td>Comparable with</td>
<td>Probable</td>
</tr>
<tr>
<td>Compatible with</td>
<td>Suspect(ed)</td>
</tr>
<tr>
<td>Consistent with</td>
<td>Suspicious (for)</td>
</tr>
<tr>
<td>Favor(s)</td>
<td>Typical (of)</td>
</tr>
<tr>
<td>Malignant appearing</td>
<td></td>
</tr>
</tbody>
</table>

4. **Do not code** histology when described as:
   - Architecture
   - Foci; focus; focal
   - Pattern
Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, C442, 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
Head and Neck Histology Rules  
C000-C148, C300-C339, C410, C411, C442, C479  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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/endolympathic 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 carcinomas 8980 and a subtype/variant of odontogenic carcinomas
- Sarcoma 8800/3 and a subtype/variant of sarcoma
- Squamous cell carcinoma 8070 and subtype/variant of squamous cell 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 (adenocarcinoma) 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
Kidney Equivalent Terms and Definitions

C649

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

(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.

<table>
<thead>
<tr>
<th>Equivalent or Equal Terms</th>
</tr>
</thead>
</table>

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
- Majority; major; predominantly; greater than 50%
- 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 using the Solid Tumor Rules, rhabdomyosarcoma and its subtypes/variants are treated the same as all NOS and subtypes/variants.
## Kidney Equivalent Terms and Definitions

### C649

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

<table>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nephroblastoma 8960</td>
<td>Wilms tumor</td>
<td></td>
</tr>
<tr>
<td>Neuroendocrine tumor (NET) 8240</td>
<td>Carcinoid [OBS] Well-differentiated neuroendocrine tumor</td>
<td>Large cell neuroendocrine carcinoma/tumor 8013 Small cell neuroendocrine carcinoma 8041</td>
</tr>
<tr>
<td>Renal cell carcinoma NOS 8312</td>
<td>RCC Sarcomatoid carcinoma Sarcomatoid renal cell carcinoma Succinate dehydrogenase-deficient renal cell carcinoma (SDHD) Unclassified renal cell carcinoma</td>
<td>Acquired cystic disease-associated renal cell carcinoma/tubulocystic renal cell carcinoma 8316* Chromophobe renal cell carcinoma (ChRCC) 8317 Clear cell papillary renal cell carcinoma 8323/3 Note: The 2016 WHO 4th Edition Classification of Tumors of the Urinary System and Male Genital Organs has reclassified this histology as a /1 because it is low nuclear grade and is now thought to be a neoplasia. This change was not implemented in the 2018 ICD-O update. Clear cell renal cell carcinoma (ccRCC) 8310 Collecting duct carcinoma 8319 Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma 8311* MiT family translocation renal cell carcinomas 8311* Note: Hereditary leiomyomatosis and renal cell carcinoma-associated renal cell carcinoma and MiT family translocation 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. Mucinous tubular and spindle cell carcinoma 8480* Papillary renal cell carcinoma (PRCC) 8260 Renal medullary carcinoma 8510* Note: This is a new term (previously called renal spindle cell carcinoma).</td>
</tr>
</tbody>
</table>

### Notes

1. WHO, IARC, and CAP agree that sarcomatoid carcinoma is a pattern of differentiation, not a specific subtype, of renal cell carcinoma.

2. Sarcomatoid is listed in the CAP Kidney protocol under the header “features.”

---

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)
<table>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sarcoma 8800/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> Rhabdomyosarcoma is a NOS with the following subtype/variants:</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alveolar rhabdomyosarcoma 8920</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Embryonal rhabdomyosarcoma 8910</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pleomorphic rhabdomyosarcoma 8901</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spindle cell/sclerosing rhabdomyosarcoma 8912</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Angiosarcoma 9120/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Clear cell sarcoma/bone-metastasizing renal tumor of childhood 8964/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leiomyosarcoma/renal vein leiomyosarcoma 8890/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Osteosarcoma 9180/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primitive/peripheral neuroectodermal tumor (pNET)/Ewing sarcoma 9364/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rhabdomyosarcoma 8900/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alveolar rhabdomyosarcoma 8920/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Embryonal rhabdomyosarcoma 8910/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pleomorphic rhabdomyosarcoma 8901/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spindle cell/sclerosing rhabdomyosarcoma 8912/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Synovial sarcoma 9040/3</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* These new codes were approved by the IARC/WHO Committee for ICD-O.
Table 2: Neoplasms which are Not Reportable

<table>
<thead>
<tr>
<th>Not Reportable Histology Term and Code</th>
<th>Synonyms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adult cystic teratoma 8959/0</td>
<td>Mixed epithelial and stromal tumor</td>
</tr>
<tr>
<td>Angiomyolipoma 8860/0</td>
<td>Renal epithelial stromal tumor</td>
</tr>
<tr>
<td>Congenital mesoblastic nephroma 8960/1</td>
<td></td>
</tr>
<tr>
<td>Cystic partially-differentiated nephroblastoma 8959/1</td>
<td>CMN</td>
</tr>
<tr>
<td>Epithelioid angiolipoma 8860/1*</td>
<td></td>
</tr>
<tr>
<td>Hemangioblastoma 9161/1</td>
<td></td>
</tr>
<tr>
<td>Hemangioma 9120/0</td>
<td></td>
</tr>
<tr>
<td>Juxtaglomerular cell tumor 8361/0</td>
<td></td>
</tr>
<tr>
<td>Leiomyoma 8890/0</td>
<td></td>
</tr>
<tr>
<td>Lymphangioma 9170/0</td>
<td></td>
</tr>
<tr>
<td>Metanephric adenofibroma 9013/0</td>
<td>Nephrogenic adenofibroma</td>
</tr>
<tr>
<td>Metanephric adenoma 8325/0</td>
<td></td>
</tr>
<tr>
<td>Metanephric stromal tumor 8935/1</td>
<td></td>
</tr>
<tr>
<td>Multilocular cystic renal neoplasm of low malignant potential 8316/1*</td>
<td></td>
</tr>
<tr>
<td>Nephrogenic rests (no code)</td>
<td></td>
</tr>
<tr>
<td>Oncocytoma 8290/0</td>
<td></td>
</tr>
<tr>
<td>Papillary adenoma 8260/0</td>
<td></td>
</tr>
<tr>
<td>Paraganglioma 8700/0</td>
<td>Extra-adrenal pheochromocytoma</td>
</tr>
<tr>
<td>Pediatric cystic nephroma 8959/0</td>
<td></td>
</tr>
<tr>
<td>Renomedullary interstitial cell tumor 8966/0</td>
<td>Medullary fibroma</td>
</tr>
<tr>
<td>Schwannoma 9560/0</td>
<td></td>
</tr>
<tr>
<td>Solitary fibrous tumor 8815/1</td>
<td></td>
</tr>
</tbody>
</table>

* 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)

A.D.A.M. illustration used with licensed permission. All rights reserved.
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**\(^1\) 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.*

\(^1\) 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 (CXX), third (CXX) and/or fourth characters (CXX).

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\(^4\) 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\(^4\) when separate/non-contiguous tumors are two or more different subtypes/variants in Column 3, Table 1 in the Equivalent Terms and Definitions.

*Note:* 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.

Rule M8  Abstract a single primary\(^1\) 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) 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)
Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M9  Abstract multiple primaries\(^4\) 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\(^1\) 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\(^1\) (the invasive) when an invasive tumor is diagnosed less than or equal to 60 days after an in situ tumor in the same kidney.

Note 1: The rules are hierarchical. Only use this rule when none of the previous rules apply.
Note 2: The tumors may be a NOS and a subtype/variant of that NOS.
Note 3: When the case has been abstracted, change behavior code on original abstract from /2 to /3.
Note 4: Do not change date of diagnosis.
Note 5: If the case has already been submitted to the central registry, report all changes.
Note 6: The physician may stage both tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).
Note 7: See the COC and SEER manuals for instructions on coding other data items such as Date of Diagnosis, Accession Year and Sequence Number.

Rule M12  Abstract multiple primaries\(^4\) when an invasive tumor occurs more than 60 days after an in situ tumor.

Note 1: The rules are hierarchical. Only use this rule when none of the previous rules apply.
Note 2: Abstract both the invasive and in situ tumors.
Note 3: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression.
Kidney Multiple Primary Rules
C649
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M13  Abstract a single primary\(^1\) 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.

\(^1\) 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.

\(^i\) Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.
IMPORTANT NOTES

   Note 1: Histology changes do occur following immunotherapy, chemotherapy, and radiation therapy.  
   Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

2. Code the histology assigned by the physician. 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. Cytology (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.

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

(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
         o Behavior unknown/not documented (use staging form to determine behavior when available)
         o Invasive
      • 8257/3 when
         o Microinvasive
         o Minimally invasive
      • 8253/2 when
         o Preinvasive
         o In situ
   
   Note: 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
         o Microinvasive
         o Minimally invasive
      • 8250/2 when
         o Preinvasive
         o 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
  o 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
  o These terms are used ONLY to determine multiple primaries
  o 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”

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

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**
### Terminology

<table>
<thead>
<tr>
<th>Terminology</th>
<th>Laterality</th>
<th>Site Term and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Bronchus intermedius</strong></td>
<td>Bilateral</td>
<td>Mainstem bronchus C340</td>
</tr>
<tr>
<td>Carina</td>
<td></td>
<td><strong>Note</strong>: 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</td>
</tr>
<tr>
<td>Hilus of lung</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Perihilar</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lingula of lung</td>
<td>Left</td>
<td>Upper lobe C341</td>
</tr>
<tr>
<td>Apex</td>
<td>Bilateral</td>
<td>Upper lobe C341</td>
</tr>
<tr>
<td>Apex of lung</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lung apex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pancoast tumor</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superior lobar bronchus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Upper lobe bronchi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Middle lobe</td>
<td>Right</td>
<td>Middle lobe C342</td>
</tr>
<tr>
<td>Middle lobe bronchi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Base of lung</td>
<td>Bilateral</td>
<td>Lower lobe C343</td>
</tr>
<tr>
<td>Lower lobar bronchus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lower lobe</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lower lobe bronchi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lower lobe segmental bronchi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Overlapping lesion of lung</td>
<td>Bilateral</td>
<td>Overlapping lesion of lung C348</td>
</tr>
<tr>
<td></td>
<td></td>
<td><strong>Note</strong>: One lesion/tumor which overlaps two or more lobes</td>
</tr>
</tbody>
</table>

Table continues on next page
Lung Equivalent Terms and Definitions  
C340-C343, C348, C349  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

<table>
<thead>
<tr>
<th>Required Terms</th>
<th>Combination Histologies and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenocarcinoma NOS AND Squamous cell carcinoma NOS</td>
<td>Adenosquamous carcinoma 8560</td>
</tr>
<tr>
<td><strong>Note:</strong> Diagnosis <strong>must be</strong> adenocarcinoma NOS and squamous cell carcinoma NOS, <strong>NOT</strong> any of the <strong>subtypes/variants</strong> of adenocarcinoma or squamous cell carcinoma</td>
<td></td>
</tr>
<tr>
<td>Giant cell carcinoma AND Spindle cell carcinoma</td>
<td>Sarcomatoid carcinoma 8033</td>
</tr>
<tr>
<td><strong>Note:</strong> 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.</td>
<td></td>
</tr>
<tr>
<td>Epithelial carcinoma AND Myoepithelial carcinoma</td>
<td>Epithelial-myoepithelial carcinoma 8562</td>
</tr>
<tr>
<td>Mucinous carcinoma, invasive AND Non-mucinous carcinoma, invasive</td>
<td>Mixed invasive mucinous and non-mucinous carcinoma 8254/3*</td>
</tr>
</tbody>
</table>
### Required Terms

- Small cell carcinoma/neuroendocrine tumor (NET)
  
  **Note:** Includes subtypes/variants of small cell/neuroendocrine tumor. See [Table 3](#) for subtypes/variants.

  AND

- At least one of the following:
  - Adenocarcinoma and any subtype/variant of adenocarcinoma
  - Adenosquamous carcinoma
  - Large cell carcinoma and any subtype/variant of large cell carcinoma
  - Squamous cell carcinoma and any subtype/variant of squamous cell carcinoma
  - Non-small cell carcinoma

### Combination Histologies and Code

- Combined small cell carcinoma 8045

### Squamous Cell Carcinoma (Epidermoid Carcinoma)

- AND

### Large Cell Non-Keratinizing Squamous Cell Carcinoma

**Note:** Squamous cell carcinoma and epidermoid carcinoma are synonyms

### Squamous Cell Carcinoma (Epidermoid Carcinoma)

- AND

### Small Cell Non-Keratinizing Squamous Cell Carcinoma

**Note:** Squamous cell carcinoma and epidermoid carcinoma are synonyms
<table>
<thead>
<tr>
<th>Required Terms</th>
<th>Combination Histologies and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Squamous cell carcinoma, keratinizing</td>
<td>Squamous cell carcinoma, NOS 8070</td>
</tr>
<tr>
<td><strong>AND</strong></td>
<td></td>
</tr>
<tr>
<td>Squamous cell carcinoma, non-keratinizing</td>
<td></td>
</tr>
<tr>
<td><strong>Squamous cell (epidermoid) carcinoma</strong></td>
<td>Squamous cell carcinoma, sarcomatoid 8074</td>
</tr>
<tr>
<td><strong>AND</strong></td>
<td>Squamous cell carcinoma, spindle cell 8074</td>
</tr>
<tr>
<td><strong>One or both</strong> of the following:</td>
<td></td>
</tr>
<tr>
<td>• Sarcomatoid carcinoma</td>
<td></td>
</tr>
<tr>
<td>• Spindle cell carcinoma</td>
<td></td>
</tr>
</tbody>
</table>

**Note 1:** Does not include subtypes/variants of squamous cell. See Table 3 for subtypes/variants.

**Note 2:** Squamous cell carcinoma and epidermoid carcinoma are synonyms.

Table continues on next page
### Required Terms

Diagnosis must be a single tumor which meets one of the following two criteria:

1. **At least two of the subtypes/variants of adenocarcinoma AND percentages of each type are unknown/not stated**
   - Acinar adenocarcinoma
   - Clear cell adenocarcinoma
   - Lepidic adenocarcinoma  
     *Note:* Lepidic adenocarcinoma may or may not have mucinous components.
   - Micropapillary adenocarcinoma
   - Papillary adenocarcinoma
   - Solid adenocarcinoma
   - Well-differentiated fetal adenocarcinoma  
     *Note:* This includes a diagnosis of adenocarcinoma AND at least two subtypes/variants of adenocarcinoma.

2. A combination of histologies **not listed on previous rows** of this table.

### Combination Histologies and Code

| Adenocarcinoma with mixed subtypes **8255/3**  
| **Note 1:** 8255 is a “last resort” code.  
| **Note 2:** See the **Histology Rules** to determine when it is appropriate to use this code for combination histologies other than adenocarcinoma subtypes/variants.  
| **Note 3:** 8255 does not apply to squamous cell carcinoma, NOS and/or subtype/variants of SCC. |
Lung Equivalent Terms and Definitions  
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.

Table begins on next page
<table>
<thead>
<tr>
<th>Specific or NOS Histology Term and Code</th>
<th>Synonym of Specific or NOS</th>
<th>Subtype/variant of NOS and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Adenocarcinoma 8140</strong></td>
<td>Adenocarcinoma NOS</td>
<td>Acinar adenocarcinoma/adenocarcinoma, acinar predominant <em>(for lung only)</em> 8551*</td>
</tr>
<tr>
<td></td>
<td>Adenocarcinoma in situ</td>
<td>Adenoid cystic/adenocystic carcinoma 8200</td>
</tr>
<tr>
<td></td>
<td>8140/2</td>
<td>Colloid adenocarcinoma 8480</td>
</tr>
<tr>
<td></td>
<td>Adenocarcinoma invasive</td>
<td>Fetal adenocarcinoma 8333</td>
</tr>
<tr>
<td></td>
<td>8140/3</td>
<td>Lepidic adenocarcinoma/adenocarcinoma, lepidic predominant 8250/3*</td>
</tr>
<tr>
<td></td>
<td>Adenocarcinoma, non-</td>
<td>Micinous carcinoma/adenocarcinoma</td>
</tr>
<tr>
<td></td>
<td>mucinous, NOS</td>
<td><em>(for lung only)</em></td>
</tr>
<tr>
<td></td>
<td></td>
<td>in situ 8253/2*</td>
</tr>
<tr>
<td>Note 1: Mucinous adenocarcinoma for</td>
<td></td>
<td>invasive 8253/3*</td>
</tr>
<tr>
<td>lung only is coded as follows:</td>
<td></td>
<td>minimally invasive 8257/3*</td>
</tr>
<tr>
<td>• 8253/3* when</td>
<td></td>
<td>microinvasive 8257/3*</td>
</tr>
<tr>
<td>o Behavior unknown/not documented</td>
<td></td>
<td>preinvasive 8253/2*</td>
</tr>
<tr>
<td>(use staging form to determine</td>
<td></td>
<td>Micropapillary adenocarcinoma</td>
</tr>
<tr>
<td>behavior when available)</td>
<td></td>
<td>adenocarcinoma, micropapillary</td>
</tr>
<tr>
<td>o Invasive</td>
<td></td>
<td>predominant 8265</td>
</tr>
<tr>
<td>• 8257/3* when</td>
<td></td>
<td>Mixed invasive mucinous and</td>
</tr>
<tr>
<td>o Microinvasive</td>
<td></td>
<td>non-mucinous adenocarcinoma 8254*</td>
</tr>
<tr>
<td>o Minimally invasive</td>
<td></td>
<td>Non-mucinous adenocarcinoma</td>
</tr>
<tr>
<td>• 8253/2* when</td>
<td></td>
<td><em>(for lung only)</em></td>
</tr>
<tr>
<td>o Preinvasive</td>
<td></td>
<td>in situ 8250/2*</td>
</tr>
<tr>
<td>o In situ</td>
<td></td>
<td>microinvasive 8250/2*</td>
</tr>
<tr>
<td>Note 2: Non-mucinous adenocarcinoma</td>
<td></td>
<td>minimally invasive 8256/3*</td>
</tr>
<tr>
<td>for lung only is coded as follows:</td>
<td></td>
<td>preinvasive 8256/2*</td>
</tr>
<tr>
<td>• 8256/3* when</td>
<td></td>
<td>Papillary adenocarcinoma/adenocarcinoma,</td>
</tr>
<tr>
<td>o Microinvasive</td>
<td></td>
<td>papillary predominant 8260</td>
</tr>
<tr>
<td>o Minimally invasive</td>
<td></td>
<td>Pulmonary intestinal-type</td>
</tr>
<tr>
<td>• 8250/2* when</td>
<td></td>
<td>adenocarcinoma/enteric adenocarcinoma 8144</td>
</tr>
<tr>
<td>o Preinvasive</td>
<td></td>
<td>Solid adenocarcinoma/adenocarcinoma, solid predominant 8230</td>
</tr>
<tr>
<td>o In situ</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Lung Equivalent Terms and Definitions

**C340-C343, C348, C349**

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

<table>
<thead>
<tr>
<th>Specific or NOS Histology Term and Code</th>
<th>Synonym of Specific or NOS</th>
<th>Subtype/variant of NOS and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Adenosquamous carcinoma 8560</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Epithelial-myoepithelial carcinoma 8562</td>
<td>Adenomyoepithelioma*</td>
<td></td>
</tr>
<tr>
<td>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</td>
<td>Epimyoepithelial carcinoma</td>
<td>Epithelial-myoepithelial tumor of unproven malignant potential*</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malignant mixed tumor comprising epithelial and myoepithelial cells</td>
</tr>
<tr>
<td>Epithelioid hemangioepithelioma 9133</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Giant cell carcinoma 8031</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intrapulmonary thymoma (arising within lung) 8580/3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Note: Intrapulmonary thymoma is always malignant /3.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table continues on next page
### Lung Equivalent Terms and Definitions

**C340-C343, C348, C349**

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

<table>
<thead>
<tr>
<th>Specific or NOS Histology Term and Code</th>
<th>Synonym of Specific or NOS</th>
<th>Subtype/variant of NOS and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Large cell carcinoma 8012</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 1:</strong> A diagnosis of large cell carcinoma is usually followed by further diagnostic testing to identify the subtype/variant.</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 2:</strong> 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.</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 3:</strong> Large cell carcinoma with neuroendocrine (NE) differentiation lacks NE morphology and is coded as large cell carcinoma, not large cell neuroendocrine carcinoma.</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Lymphoepithelioma-like carcinoma 8082</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Melanoma 8720</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Mucoepidermoid carcinoma 8430</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> Mucoepidermoid tumor is listed as a synonym of mucoepidermoid carcinoma in WHO</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Myoepithelial carcinoma 8982</strong></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Lung Equivalent Terms and Definitions

C340-C343, C348, C349

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

<table>
<thead>
<tr>
<th>Specific or NOS Histology Term and Code</th>
<th>Synonym of Specific or NOS</th>
<th>Subtype/variant of NOS and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>NUT carcinoma 8023/3</strong>&lt;br&gt;NUT: nuclear protein in tests&lt;br&gt;NUT/M1 gene rearrangement</td>
<td>Aggressive t(15:19) positive carcinoma&lt;br&gt;BET-rearranged carcinoma&lt;br&gt;Carcinoma with t(15:19) translocation&lt;br&gt;Midline carcinoma of children and young adults with NUT rearrangement&lt;br&gt;Midline lethal carcinoma&lt;br&gt;NUT midline carcinoma</td>
<td></td>
</tr>
<tr>
<td><strong>PEComa malignant 8714/3</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Note: Tumor displays perivascular epithelioid (PEC) differentiation</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Pleomorphic carcinoma 8022</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>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</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Note 2: Pleomorphic carcinoma has components of adenocarcinoma and/or large cell carcinoma, also squamous carcinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Specific or NOS Histology Term and Code</td>
<td>Synonym of Specific or NOS</td>
<td>Subtype/variant of NOS and Code</td>
</tr>
<tr>
<td>----------------------------------------</td>
<td>---------------------------</td>
<td>--------------------------------</td>
</tr>
<tr>
<td>Sarcoma NOS 8800/3</td>
<td></td>
<td>Biphasic synovial sarcoma 9043/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Epithelioid cell synovial sarcoma 9042/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pulmonary artery intimal sarcoma/low-grade malignant myxoid endobronchial tumor 9173/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Pulmonary myxoid sarcoma with EWSR1 - CREB1 translocation 8842/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Spindle cell synovial sarcoma 9041/3</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Synovial sarcoma 9040/3</td>
</tr>
<tr>
<td>Small cell carcinoma 8041/3</td>
<td>Reserve cell carcinoma</td>
<td>Atypical carcinoid 8249/3</td>
</tr>
<tr>
<td></td>
<td>Round cell carcinoma</td>
<td>Combined small cell carcinoma 8045/3</td>
</tr>
<tr>
<td></td>
<td>SCLC</td>
<td>Large cell neuroendocrine carcinoma/combined large cell neuroendocrine carcinoma 8013/3</td>
</tr>
<tr>
<td></td>
<td>Small cell carcinoma NOS</td>
<td>Typical carcinoid 8240/3</td>
</tr>
<tr>
<td></td>
<td>Small cell neuroendocrine carcinoma</td>
<td>Neuroendocrine carcinoma, NOS</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Well-differentiated neuroendocrine carcinoma</td>
</tr>
<tr>
<td>Spindle cell carcinoma 8032</td>
<td>Epidermoid carcinoma NOS</td>
<td>Basaloid carcinoma/basaloid squamous cell carcinoma 8083</td>
</tr>
<tr>
<td></td>
<td>Squamous cell carcinoma</td>
<td>Keratinizing squamous cell carcinoma 8071</td>
</tr>
<tr>
<td></td>
<td>Epithelioma</td>
<td>Non-keratinizing carcinoma 8072</td>
</tr>
</tbody>
</table>

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

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)
Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Illustrations

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

Mediastinum
Used with permission

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

Lymph Nodes Lung
Used with permission
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
Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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**† 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

† 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 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\(^i\) when there are separate, non-contiguous tumors in sites with ICD-O site codes that differ at the second CXxx and/or third character CxXx.

*Note:* When codes differ at the second or third characters, the tumors are in different primary sites.

Rule M4 Abstract multiple primaries\(^i\) 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 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** 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** 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:** 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 8520/3 is a subtype of adenocarcinoma 8140/3. They are distinctly different histologies. Abstract multiple primaries.

Rule M7  Abstract a **single primary** 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 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 8250/3. 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 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)

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

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

Rule M10 Abstract a single primary\textsuperscript{1} when an in situ tumor is diagnosed after an invasive tumor AND tumors occur in the same lung.

\textit{Note 1:} The rules are hierarchical. Only use this rule when none of the previous rules apply.

\textit{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.

\textit{Note 3:} The in situ is recorded as a recurrence for those registrars who collect recurrence data.

Rule M11 Abstract multiple primaries\textsuperscript{2} when there is a single tumor in each lung (one tumor in the right lung and one tumor in the left lung).

\textit{Note 1:} The only exception is when there is proof\textsuperscript{3} 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)

\textit{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).

\textit{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.

\textit{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.

\textit{Note 5:} Lymph node involvement is recorded in staging criteria.
Lung Multiple Primary Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M12  Abstract a single primary\textsuperscript{1} (the invasive) when an invasive tumor is diagnosed less than or equal to 60 days after an in situ tumor in the same lung.

\textit{Note 1:} The rules are hierarchical. Only use this rule when none of the previous rules apply.

\textit{Note 2:} The tumors may be a NOS and a subtype/variant of that NOS.

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

\textit{Note 4:} Do not change date of diagnosis.

\textit{Note 5:} If the case has already been submitted to the central registry, report all changes.

\textit{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).

\textit{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\textsuperscript{4} when an invasive tumor occurs more than 60 days after an in situ tumor in the same lung.

\textit{Note 1:} The rules are hierarchical. Only use this rule when none of the previous rules apply.

\textit{Note 2:} Abstract both the invasive and in situ tumors.

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

\textit{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\textsuperscript{1} when none of the previous rules apply.

\textit{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

---

\textsuperscript{1} 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.

\textsuperscript{4} 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**

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

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

*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 site often shows variations from the primary tumor. When it is the only tissue available, it is more accurate than a scan.

4. **Scan:** The following list is 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 8510. 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/documentated
   
   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
- Appears
- Comparable with
- Compatible with
- Consistent with
- Favor(s)
- Malignant appearing
- Most likely
- Presumed
- Probable
- Suspect(ed)
- Suspicious (for)
- Typical (of)
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.
Lung Histology Rules  
C340-C343, C348, C349  
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**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
- The case is accessioned (added to your database) based on a single histology described by ambiguous terminology and no other histology information is available/documentated

*Note:* If the case does not meet the criteria in the first two bullets, code non-small cell lung cancer (NSCLC) **8046**.

**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 bullet 3.
**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, 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:** 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:* 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 are **new codes and terms** which will allow mucinous adenocarcinoma/carcinoma 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 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.
Lung Histology Rules
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

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.**

---

Jump to [Equivalent Terms and Definitions](#)  
Jump to [Multiple Primary Rules](#)
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.

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.
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
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)

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).
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)

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:

1. The behavior must be malignant:
   A. Pathology designates the behavior as malignant/invasive, 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 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)
Information is presented in the general order in which a case is abstracted.

**Section 1: Behavior Code**
A. **Priority Order** for Using Documentation to Assign Behavior
B. **Table 1**: WHO Grades for Select CNS Neoplasms

**Section 2: Reportable Primary Sites and Histologies**
A. **Priorities for Coding Primary Site**
B. **Reportable Primary Site Groups**
C. **Table 2**: Reportable Primary Sites
D. **Table 3**: Specific Histologies, NOS, and Subtypes/Variants
E. **Table 4**: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves

**Section 3: Additional information to complete the abstract**
A. **Conflicting information on Pathology report(s)**
B. **Table 5**: Paired Sites
C. **Table 6**: Non-malignant CNS Tumors with the Potential of Transforming to Malignant Behavior
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. Scan, use behavior information from radiography in the following priority order:
   A. MRI
   B. CT
   C. PET
   D. Angiogram
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)

6. When instructions 1-5 **do not apply**, use **Table 1** to determine behavior.

### Table 1: WHO Grades for Select CNS Neoplasms

<table>
<thead>
<tr>
<th>WHO Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>WHO Grade I</td>
<td>Circumscribed tumors of low proliferative potential associated with the possibility of cure following resection</td>
</tr>
<tr>
<td>WHO Grade II</td>
<td>Infiltrative tumors with low proliferative potential with increased risk of recurrence</td>
</tr>
<tr>
<td>WHO Grade III</td>
<td>Tumors with histologic evidence of malignancy, including nuclear atypia and mitotic activity, associated with an aggressive clinical course</td>
</tr>
<tr>
<td>WHO Grade IV</td>
<td>Tumors that are cytologically malignant, mitotically active, and associated with rapid clinical progression and potential for dissemination</td>
</tr>
</tbody>
</table>

**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

**Table Instructions**

1. Use the **malignant CNS** rules for all WHO Grade 3 and 4.
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 1** (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 and WHO Grade

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaplastic (malignant) meningioma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic astrocytoma, IDH-mutant</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic ependymoma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic ganglioglioma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic oligodendroglioma IDH-mutant and 1p/19q-codeleted</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic pleomorphic xanthroastrocytoma</td>
<td>3</td>
</tr>
<tr>
<td>Angiocentric glioma</td>
<td>1</td>
</tr>
<tr>
<td>Atypical choroid plexus papilloma</td>
<td>2</td>
</tr>
<tr>
<td>Atypical meningioma</td>
<td>2</td>
</tr>
<tr>
<td>Atypical teratoid/rhabdoid tumor</td>
<td>4</td>
</tr>
<tr>
<td>Central neurocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Cerebellar liponeurocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Chordoid glioma of third ventricle</td>
<td>2</td>
</tr>
<tr>
<td>Choroid plexus carcinoma</td>
<td>3</td>
</tr>
<tr>
<td>Choroid plexus papilloma</td>
<td>1</td>
</tr>
<tr>
<td>CNS embryonal tumor NOS</td>
<td>4</td>
</tr>
<tr>
<td>CNS embryonal tumor with rhabdoid features</td>
<td>4</td>
</tr>
<tr>
<td>Craniopharyngioma</td>
<td>1</td>
</tr>
<tr>
<td>Desmoplastic infantile astrocytoma and ganglioglioma</td>
<td>1</td>
</tr>
<tr>
<td>Diffuse astrocytoma, IDH-mutant</td>
<td>2</td>
</tr>
<tr>
<td>Diffuse midline glioma, H3K27M-mutant</td>
<td>4</td>
</tr>
<tr>
<td>Dysembryoplastic neuroepithelial tumor</td>
<td>1</td>
</tr>
<tr>
<td>Dysplastic gangliocytoma of cerebellum (Lhemitte-Duclos)</td>
<td>1</td>
</tr>
<tr>
<td>Ependymoma</td>
<td>2</td>
</tr>
</tbody>
</table>
## Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

**C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**

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

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ependymoma, RELA fusion-positive</td>
<td>2 or 3</td>
</tr>
<tr>
<td><strong>Note:</strong> Tissue/pathology reports or CAP protocol/summary will specify whether this is WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Extraventricular neurocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Gangliocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Glioblastoma, IDH-mutant</td>
<td>4</td>
</tr>
<tr>
<td>Glioblastoma, IDH-wildtype</td>
<td>4</td>
</tr>
<tr>
<td>Granular cell tumor</td>
<td>1</td>
</tr>
<tr>
<td>Hemangioblastoma</td>
<td>1</td>
</tr>
<tr>
<td>Malignant peripheral nerve sheath tumor (MPNST)</td>
<td>2, 3, or 4</td>
</tr>
<tr>
<td><strong>Note:</strong> Tissue/pathology reports or CAP protocol/summary will specify whether this is WHO Grade 2, 3, or 4</td>
<td></td>
</tr>
<tr>
<td>Medulloblastoma (including all subtypes)</td>
<td>4</td>
</tr>
<tr>
<td>Medulloepithelioma</td>
<td>4</td>
</tr>
<tr>
<td>Meningioma</td>
<td>1</td>
</tr>
<tr>
<td>Myxopapillary ependymoma</td>
<td>1</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td>1</td>
</tr>
<tr>
<td>Oligodendroglioma IDH-mutant and 1p/19q deleted</td>
<td>2</td>
</tr>
<tr>
<td>Papillary glioneuronal tumor</td>
<td>1</td>
</tr>
<tr>
<td>Papillary tumor of the pineal region</td>
<td>2 or 3</td>
</tr>
<tr>
<td><strong>Note:</strong> Tissue/pathology reports or CAP protocol/summary will specify whether it is WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Perineuroma</td>
<td>1</td>
</tr>
<tr>
<td>Pilocytic astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td><strong>Note:</strong> Collected as malignant /3 in North America</td>
<td></td>
</tr>
</tbody>
</table>
### Histology

<table>
<thead>
<tr>
<th>Tissue/Pathology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pineal parenchymal tumor of intermediate differentiation</td>
<td>2 or 3</td>
</tr>
<tr>
<td>Note: Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Pineoblastoma</td>
<td>4</td>
</tr>
<tr>
<td>Pineocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Pituicytoma</td>
<td>1</td>
</tr>
<tr>
<td>Pleomorphic xanthroastrocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Rosette-forming glioneuronal tumor</td>
<td>1</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>1</td>
</tr>
<tr>
<td>Solitary fibrous tumor/hemangiopericytoma</td>
<td>1, 2, or 3</td>
</tr>
<tr>
<td>Note: Tissue/pathology reports or CAP protocol/summary will specify WHO Grade 1, 2, or 3</td>
<td></td>
</tr>
<tr>
<td>Spindle cell oncocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Subependymal giant cell astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Subependymoma</td>
<td>1</td>
</tr>
</tbody>
</table>
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
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).
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
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)

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
Use Table 2 to **determine** whether a primary site is **reportable**.

<table>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brain</td>
<td>Brain NOS C719</td>
</tr>
<tr>
<td></td>
<td>Brain stem C717</td>
</tr>
<tr>
<td></td>
<td>Cerebellum NOS C716</td>
</tr>
<tr>
<td></td>
<td>Cerebrum C710</td>
</tr>
<tr>
<td></td>
<td>Frontal lobe C711</td>
</tr>
<tr>
<td></td>
<td>Occipital lobe C714</td>
</tr>
<tr>
<td></td>
<td>Overlapping lesion of brain C718</td>
</tr>
<tr>
<td></td>
<td>Parietal lobe C713</td>
</tr>
<tr>
<td></td>
<td>Temporal lobe C712</td>
</tr>
<tr>
<td></td>
<td>Ventricle NOS C715</td>
</tr>
<tr>
<td>Cranial Nerves</td>
<td>Abducent (cranial nerve VI) C725</td>
</tr>
<tr>
<td></td>
<td>Accessory (cranial nerve XI) C725</td>
</tr>
<tr>
<td></td>
<td>Acoustic (cranial nerve VIII) C724</td>
</tr>
<tr>
<td></td>
<td>Cranial nerve NOS C725</td>
</tr>
<tr>
<td></td>
<td>Facial (cranial nerve VII) C725</td>
</tr>
<tr>
<td></td>
<td>Glossopharyngeal (cranial nerve IX) C725</td>
</tr>
<tr>
<td></td>
<td>Hypoglossal (cranial nerve XII) C725</td>
</tr>
<tr>
<td></td>
<td>Oculomotor (cranial nerve III) C725</td>
</tr>
<tr>
<td></td>
<td>Olfactory (cranial nerve I) C722</td>
</tr>
<tr>
<td></td>
<td>Optic (cranial nerve II) C723</td>
</tr>
<tr>
<td></td>
<td>Trigeminal (cranial nerve V) C725</td>
</tr>
<tr>
<td></td>
<td>Trochlear (cranial nerve IV) C725</td>
</tr>
<tr>
<td></td>
<td>Vagus (cranial nerve X) C725</td>
</tr>
</tbody>
</table>
### Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

#### Site Group

<table>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
</table>
| Ill-Defined Sites Central Nervous System | Nervous system NOS C729  
Overlapping lesion of brain and central nervous system C728 |
| Intracranial Duct and Glands       | Craniopharyngeal duct C752  
Pineal gland C753  
Pituitary gland C751 |
| Meninges                           | Cerebral meninges C700  
Meninges NOS C709  
Spinal meninges C701 |
| Peripheral Nerve and Autonomic Nervous System | Abdomen C474  
Autonomic nervous system NOS C479  
Head, face and neck C470  
Lower limb and hip C472  
Overlapping lesion of peripheral nerves and autonomic nervous system C478  
Thorax C473  
Trunk NOS C476  
Upper limbs and shoulder C471  
Spinal Nerve NOS C479 |
| Spinal Sites                        | Cauda equina/conus medullaris/filum terminale C721  
Meninges NOS C709  
Spinal cord C720  
Spinal meninges C701 |
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 using the Solid Tumor Rules, chondrosarcoma and mesenchymal chondrosarcoma are treated the same as all NOS and subtypes/variants.

**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)

<table>
<thead>
<tr>
<th>Specific and NOS Histology Codes</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaplastic ganglioglioma 9505</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Astroblastoma 9430</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
| Astrocytoma NOS 9400             | Diffuse astrocytoma IDH-mutant  
Diffuse astrocytoma IDH-wildtype  
Diffuse astrocytoma NOS | Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS 9401  
Gemistocytic astrocytoma IDH-mutant 9411  
Pleomorphic xanthroastrocytoma /anaplastic pleomorphic xanthroastrocytoma 9424 |
| Choriocarcinoma 9100             |          |                   |
| Choroid plexus carcinoma 9390    |          |                   |
| CNS embryonal tumor with rhabdoid features 9508 | Atypical teratoid/rhabdoid tumor  
Embryonal tumor with rhabdoid features | CNS embryonal tumor 9473 |
| CNS ganglioneuroblastoma 9490    |          | CNS embryonal tumor 9473 |
| CNS neuroblastoma 9500           |          | CNS embryonal tumor 9473 |
| Diffuse midline glioma H3 K27M mutant 9385* | CNS embryonal tumor 9473 |
| Embryonal carcinoma 9070         |          | Yolk sac tumor 9071 |
| Embryonal tumor with multilayered rosettes C19MC-altered 9478* | Embryonal tumor with multilayered rosettes, NOS  
ETMR | CNS embryonal tumor 9473 |
| Ependymoma 9391                  | Clear cell ependymoma  
Tanyctytic ependymoma | Anaplastic ependymoma 9392  
Ependymoma, RELA fusion-positive 9396*  
Papillary ependymoma 9393 |
<p>| Epithelioid hemangioendothelioma 9133 |          |                   |
| Germinoma 9064                   |          |                   |</p>
<table>
<thead>
<tr>
<th>Specific and NOS Histology Codes</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Glioblastoma NOS 9440          | Glioblastoma multiforme  
GBM  
Glioblastoma, IDH wild-type  
Epithelioid glioblastoma | Giant cell glioblastoma 9441  
Glioblastoma IDH-mutant 9445*  
Gliosarcoma 9442 |
| Immature teratoma 9080         |          | Mixed germ cell tumor 9085  
Teratoma with malignant transformation 9084 |
| Malignant meningioma 9530      | Anaplastic meningioma | Papillary/rhabdoid meningioma 9538 |
| Malignant peripheral nerve sheath tumor 9540 | Epithelioid malignant peripheral nerve sheath tumor  
Malignant perineurioma  
MPNST  
MPNST with perineural differentiation | |
| Medulloblastoma NOS 9470       | Classic medulloblastoma | Anaplastic/large cell medulloblastoma 9474  
Medulloblastoma described as one of the following 9471  
Desmoplastic  
SHH-activated and TP53-wildtype  
With extensive nodularity  
Nodular  
Medulloblastoma non-WNT/non-SHH; medulloblastoma group 3 or group 4 9477*  
Medulloblastoma SHH-activated and TP53-mutant 9476*  
Medulloblastoma WNT-activated 9475* |
| Medulloepithelioma 9501        |          | |
| Meningeal melanoma 8720        |          | Meningeal melanomatosis 8728 |
| Oligoastrocytoma NOS 9382      | Anaplastic oligoastrocytoma NOS | |
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>
<thead>
<tr>
<th>Specific and NOS Histology Codes</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oligodendroglioma NOS 9450</td>
<td>Oligodendroglioma 1p/19q-codeleted Oligodendroglioma IDH-mutant Oligodendroglioma IDH-mutant and 1p/19q-codeleted</td>
<td>Anaplastic oligodendroglioma NOS 9451 IDH-mutant 1p/19q-codeleted IDH-mutant and 1p/19q-codeleted</td>
</tr>
<tr>
<td><strong>Note:</strong> Oligodendroglioma NOS is used when molecular markers cannot fully be determined</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peripheral primitive neuroectodermal tumor 9364</td>
<td>Ewing sarcoma PpNET</td>
<td>Pilomyxoid astrocytoma 9425</td>
</tr>
<tr>
<td><strong>Note:</strong> ICD-O-3 lists a behavior code of /1. It is collected as /3 in North America.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pilocytic astrocytoma 9421</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pineal parenchymal tumor of intermediate differentiation 9362</td>
<td>Pineoblastoma</td>
<td>Papillary tumor of the pineal region 9395</td>
</tr>
<tr>
<td>Sarcoma NOS 8800</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 1:</strong> Chondrosarcoma 9220 has the following subtype/variant: Mesenchymal chondrosarcoma 9240</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Note 2:</strong> Leiomyosarcoma 8890 has the following subtypes/variants: Epithelioid leiomyosarcoma 8891 Myxoid leiomyosarcoma 8896</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Solitary fibrous tumor grade 3 8815</td>
<td>Hemangiopericytoma grade 3 Solitary fibrous tumor/Hemangiopericytoma grade 3 (CNS)</td>
<td></td>
</tr>
</tbody>
</table>

* These new codes were approved by the IARC/WHO Committee for ICD-O
Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves

**Note 1:** Neoplasms arising in a **cranial** or **spinal** nerve are coded to the specific nerve in which it arises.

**Note 2:** Neoplasms, commonly meningiomas, arising in the **dura/meninges** of an **intracranial** nerve (cranial nerve within the skull) are coded to **cerebral meninges** C700.

**Note 3:** Neoplasms, commonly meningiomas, arising in the **dura/meninges** of the **spinal nerve roots** are coded to the ICD-O site code **spinal meninges** C701.

**Note 4:** It is important to check the **operative report** to determine whether the surgery is **intracranial** or **intradural**.

Use Table 4 to determine whether a malignant cranial nerve tumor should be coded to a cranial nerve or to a peripheral nerve. The anatomic location of the tumor determines whether the primary site is a peripheral or a cranial nerve.

**Column 1:** The **proper name** for the cranial nerve (CN) and the **cranial nerve number**

**Column 2:** The point at which the nerve exits the cranium

**Column 3:** Portions of the nerve coded to cranial nerve

**Column 4:** Portions of nerve coded to peripheral nerve

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cranial nerve NOS</td>
<td>Within cranium, unknown which nerve C725</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Olfactory CN 1</td>
<td>Cribriform plate</td>
<td>Surface of the brain C722</td>
<td>Originates on the olfactory mucosa of nasal cavity, then travels through the cribriform plate of the ethmoid bone C470</td>
</tr>
<tr>
<td>Optic CN 2</td>
<td>Optic canal</td>
<td>All portions are covered by meninges/dura so are reportable as C723</td>
<td></td>
</tr>
<tr>
<td>Oculomotor CN 3</td>
<td>Superior orbital fissure</td>
<td>Originates in the midbrain C725</td>
<td>After exiting the superior orbital fissure, the nerve enters the orbit C470</td>
</tr>
</tbody>
</table>
## Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions

**C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753**

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

### Table: Cranial and Peripheral Nerve Sites

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trochlear CN 4</td>
<td>Superior orbital fissure</td>
<td>Arises from the <strong>dorsal brain stem</strong>, loops around the brainstem and passes anteriorly within the <strong>subarachnoid space</strong>. It travels between the <strong>superior cerebellar and posterior cerebral arteries</strong> and through the <strong>dura</strong>, enters <strong>cavernous sinus C725</strong></td>
<td>Enters the orbital fissure <strong>C470</strong></td>
</tr>
</tbody>
</table>
| Trigeminal CN 5 | The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina.  
Superior orbital fissure.  
The foramen rotundum  
The foramen ovale. | **CN5** originates in the **pons**. Upon leaving the pons it enters a **small fossa** posterior and inferolateral to the cavernous sinus called **Meckel's (trigeminal) cave C725**. |  
**- Ophthalmic nerve branch** crosses the **pterygopalatine fossa**, inclines laterally on the back of the **maxilla**, and enters the **orbit** through the inferior orbital fissure where it is called the **infraorbital** nerve. It ends beneath the **quadatus labii superius**, and divides into multiple branches that spread to the side of the **nose**, the lower **eyelid**, and the upper **lip C470**  
**- Maxillary nerve leaves foramen rotundum and traverses the **infraorbital groove and canal** in the floor of the **orbit**, and appears on the face at the infraorbital foramen **C470**  
**- Mandibular** nerve leaves via the foramen 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)

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abducent CN 6</td>
<td>Cranial meninges</td>
<td>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</td>
<td>Dorello's canal and travels to the tip of the temporal bone. Enters orbit C470</td>
</tr>
<tr>
<td>Facial CN 7</td>
<td>Internal acoustic meatus</td>
<td>CN7 originates in the pons, along the posterior cranial fossa (posterior cranial fossa is part of the intracranial cavity.) C725</td>
<td>Enters the temple through the internal auditory meatus and runs through the facial canal. C470</td>
</tr>
</tbody>
</table>
| Acoustic or vestibulocochlear CN 8 | Internal acoustic meatus | Originates in the brain stem (medulla oblongata) between the base of the brain (pons) and the spinal cord C724  
Both the vestibular branch and the cochlear branch are located in the inner ear | |
| Glossopharyngeal CN 9          | Jugular foraminaa      | Originates in the anterior portion of the medulla oblongata C725 | Jugular foramen  
Between the internal jugular vein and internal carotid artery  
Lies on the stylopharyngeus and middle pharyngeal constrictor muscle  
Passes under the hypoglossus muscle  
Palatine tonsil  
Extends to mucous glands of the mouth, and base of the tongue C470 |

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)  
Malignant CNS Solid Tumor Rules 2018  
July 2019 Update
## 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>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Site Code: Cranial Nerve</th>
<th>Site Code: Peripheral Nerve</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vagus CN 10</td>
<td>Jugular foramen</td>
<td>The vagus nerve originates from the medulla of the brainstem. C725</td>
<td>CN10 descends within the carotid sheath medial to the internal jugular vein at the root of the neck C470. The right vagus crosses in front of the subclavian artery and travels into the fat behind the blood vessels, reaching the thorax. It then inclines behind the hilum of the right lung and moves toward the esophagus. The nerve splits into the right and left vagus at the esophageal plexus C473. Forms the anterior and posterior gastric nerves C475</td>
</tr>
<tr>
<td>Accessory CN 11</td>
<td>Jugular foramen</td>
<td>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 nerve itself C725</td>
<td>The nerve exits the skull through the jugular foramen. It then runs along the internal carotid artery within the neck C470. Reaches the sternocleidomastoid muscle and the trapezius C476</td>
</tr>
<tr>
<td>Hypoglossal CN 12</td>
<td>Hypoglossal canal</td>
<td>CN12 starts in the hypoglossal nucleus of the brainstem, C725</td>
<td>CN12 exits the hypoglossal canal, traveling between the carotid artery and jugular vein, ending under the tongue C470</td>
</tr>
</tbody>
</table>
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.
Use **Table 5** to identify sites for which laterality **must** be coded. Do **not** use this table to determine multiple primaries.

### Table 5: Paired Sites

<table>
<thead>
<tr>
<th>Paired Sites and Codes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acoustic nerve C724</td>
</tr>
<tr>
<td>Cerebral meninges C700</td>
</tr>
<tr>
<td>Cerebrum C710</td>
</tr>
<tr>
<td>Cranial nerves C725</td>
</tr>
<tr>
<td>Frontal lobe C711</td>
</tr>
<tr>
<td>Occipital lobe C714</td>
</tr>
<tr>
<td>Olfactory nerve C722</td>
</tr>
<tr>
<td>Optic nerve C723</td>
</tr>
<tr>
<td>Parietal lobe C713</td>
</tr>
<tr>
<td>Temporal lobe C712</td>
</tr>
</tbody>
</table>

**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.
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.

<table>
<thead>
<tr>
<th>Original Histology and Code</th>
<th>Transformed Histology and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chondroma 9220/0</td>
<td>Chondrosarcoma 9220/3</td>
</tr>
<tr>
<td>Ganglioglioma 9505/1</td>
<td>Anaplastic ganglioglioma 9505/3</td>
</tr>
<tr>
<td>Hemangioma 9120/0</td>
<td>Angiosarcoma 9120/3</td>
</tr>
<tr>
<td>Hemangiopericytoma 9150/1</td>
<td>Anaplastic hemangiopericytoma 9150/3</td>
</tr>
<tr>
<td>Leiomyoma 8890/0</td>
<td>Leiomyosarcoma 8890/3</td>
</tr>
<tr>
<td>Lipoma 8850/0</td>
<td>Liposarcoma 8850/3</td>
</tr>
<tr>
<td>Osteoma 9180/0</td>
<td>Osteosarcoma 9180/3</td>
</tr>
<tr>
<td>Perineurioma 9571/0</td>
<td>Malignant perineurioma 9571/3</td>
</tr>
<tr>
<td>Rhabdomyoma 8900/0</td>
<td>Rhabdomyosarcoma 8900/3</td>
</tr>
<tr>
<td>Teratoma 9080/1</td>
<td>Immature teratoma 9080/3</td>
</tr>
<tr>
<td>Teratoma, mature 9080/0</td>
<td>Immature teratoma 9080/3</td>
</tr>
</tbody>
</table>
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

A.D.A.M illustration used with licensed permission. All rights reserved.
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)

Copyright © Sinauer Associates. Licensed permission granted.
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)
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)
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)
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: 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\(^1\) 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.

\(^1\)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** 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.

**Rule M3**
Code a **single primary** 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** (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:
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)

- 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 abstract 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.

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)

<table>
<thead>
<tr>
<th>IMPORTANT: The <strong>major difference</strong> between M4 and M5 is:</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>M4:</strong> No resection as first course of treatment AND when the same tumor is subsequently resected, pathology proves malignant behavior</td>
</tr>
<tr>
<td><strong>M5:</strong> Tumor resected as first course of treatment. Subsequent tumor (recurrence or de novo) is malignant</td>
</tr>
</tbody>
</table>

**Rule M5** Abstract multiple primaries 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.
Rule M6  Abstract multiple primaries\(^\text{II}\) 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
Rule M7

Abstract a **single primary**\(^1\) 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.
Rule M8  Abstract multiple primaries\(^{\text{d}}\) when multiple tumors are present in any of the following sites or subsites:

- Any lobe of the brain \(\text{C710-C719 AND any other part of CNS}\)
- Cauda equina \(\text{C721 AND any other part of CNS}\)
- Cerebral meninges \(\text{C700 AND spinal meninges C701}\)
- Cerebral meninges \(\text{C700 AND any other part of CNS}\)
- Any one of the cranial nerves \(\text{C722-C725 AND any other part of the CNS}\)
- Any two or more of the cranial nerves
  - \(\text{C722 Olfactory, C723 Optic, C724 Acoustic, C725 Cranial nerves NOS}\)
- Meninges of cranial or peripheral nerves \(\text{C709 AND any other part of the CNS}\)
- Spinal cord \(\text{C720 AND any other part of CNS}\)
- Spinal meninges \(\text{C701 AND any other part of CNS}\)

Rule M9  Abstract multiple primaries\(^{\text{d}}\) 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\(^{\text{i}}\) 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)
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.

\(^{i}\) 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.
Note: Non-malignant CNS tumors have a separate set of rules.

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

   
   Note 1: Histology changes do occur following immunotherapy, chemotherapy and radiation therapy.
   
   Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

2. Code the histology assigned by the physician. 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.
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 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 site 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 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/document

   **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
Single Tumor

Rule H1  Code the reportable CNS tumor (Table 3 in the Equivalent Terms and Definitions) when a patient has any of the following:
  - Neurofibromatosis type 1 (NF1)
  - Neurofibromatosis type 2 (NF2)
  - Schwannomatosis

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

  Note 2: Tumors are reportable when they meet the behavior (/3) and histology requirements (see Reportability Criteria).

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

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

Rule H2  Code malignant meningioma 9530 when the diagnosis specifically states malignant/invasive.

  Note: Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.

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

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

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

  Note 3: Submit a question to Ask a SEER Registrar when the histology code is not found in Table 3, ICD-O or all updates.
Rule H4  
Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS such as the following:

*Note:* All tumors are malignant/invasive /3.
- Astrocytoma 9400 and a subtype/variant of astrocytoma
- CNS ganglioneuroblastoma 9490 and a subtype/variant of CNS ganglioneuroblastoma
- Embryonal carcinoma 9070 and a subtype/variant of embryonal carcinoma
- Ependymoma 9391 and a subtype/variant of ependymoma
- Glioblastoma 9440 and a subtype/variant of glioblastoma
- Immature teratoma 9080 and a subtype/variant of immature teratoma
- Malignant meningioma 9530 and a subtype/variant of malignant meningioma
- Malignant peripheral nerve sheath tumor 9540 and a subtype/variant of malignant peripheral nerve sheath tumor
- Medulloblastoma 9470 and a subtype/variant of medulloblastoma
- Meningeal melanoma 8720 and a subtype/variant of meningeal melanoma
- Oligodendroglioma 9450 and a subtype/variant of oligodendroglioma
- Pilocytic astrocytoma 9421 and a subtype/variant of pilocytic astrocytoma
- Pineal parenchymal tumor of intermediate differentiation 9362 and a subtype/variant of pineal parenchymal tumor of intermediate differentiation
- Sarcoma 8800 and a subtype/variant of sarcoma

*Note:* See Table 3 in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

This is the end of instructions for Single Tumor

---

Multiple Tumors Abstracted as a Single Primary

Rule H5  
Code malignant meningioma 9530 when the diagnosis specifically states malignant/invasive.

*Note:* Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.
Malignant CNS and Peripheral Nerves Histology Rules
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule H6  Code the histology when only one histology is present in all tumors.

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

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

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

Rule H7  Code the subtype/variant when all tumors are a NOS and a single subtype/variant of that NOS such as the following:

*Note:* 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 is reportable in North America as a malignant neoplasm- 9421/3.
- 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 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.
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)

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
- Majority; major; predominantly; greater than 50%
- Non-malignant is synonymous with:
  o /0 Benign
  o /1 Uncertain whether benign or malignant; borderline malignancy; low malignant potential; uncertain malignant potential
  o WHO Grade 1
- Site; topography
- Subarachnoid space; between the arachnoid mater and the pia mater of spinal meninges
- Type; subtype; variant
- Tumor; mass; tumor mass; lesion; neoplasm
  o 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
  o These terms are used ONLY for determining multiple primaries
  o DO NOT USE these terms for casefinding or determining reportability
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)

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

Section 1: Behavior Code
   A. Priority Order for Using Documentation to Assign Behavior
   B. Table 1: WHO Grades of Select CNS Neoplasms

Section 2: Reportable Primary Sites and Histologies
   A. Priorities for Coding Primary Site
   B. Reportable Primary Site
   C. Table 2: Reportable Primary Sites
   D. Table 3: Reportability of Non-Malignant Cranial Nerve (CN) Tumors (defines which portions of the cranial nerves are reportable)
   E. Table 4: Non-Reportable Neoplasms
   F. Table 5: Histologic Types of Non-Malignant Intracranial (Brain and Gland) Tumors
   G. Table 6: Specific Histologies, NOS, and Subtypes/Variants
      Note: It is important to understand that non-malignant neoplasms do occur within the brain tissue.

Section 3: Additional Information to Complete Abstract
   A. Conflicting information on Pathology report(s)
   B. Table 7: Paired Sites
   C. Table 8: Non-malignant CNS Tumors with the Potential of Transforming to Malignant Behavior
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
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)

C. PET
D. Angiogram

6. When above instructions do not apply, use Table 1 below to determine behavior.

<table>
<thead>
<tr>
<th>Table 1: WHO Grades of Select CNS Neoplasms</th>
</tr>
</thead>
<tbody>
<tr>
<td>WHO Grade</td>
</tr>
<tr>
<td>WHO Grade I</td>
</tr>
<tr>
<td>WHO Grade II</td>
</tr>
<tr>
<td>WHO Grade III</td>
</tr>
<tr>
<td>WHO Grade IV</td>
</tr>
</tbody>
</table>

**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**

<table>
<thead>
<tr>
<th>WHO Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>WHO Grade I</td>
<td>Circumscribed tumors of low proliferative potential associated with the possibility of cure following resection</td>
</tr>
<tr>
<td>WHO Grade II</td>
<td>Infiltrative tumors with low proliferative potential with increased risk of recurrence</td>
</tr>
<tr>
<td>WHO Grade III</td>
<td>Tumors with histologic evidence of malignancy, including nuclear atypia and mitotic activity, associated with an aggressive clinical course</td>
</tr>
<tr>
<td>WHO Grade IV</td>
<td>Tumors that are cytologically malignant, mitotically active, and associated with rapid clinical progression and potential for dissemination</td>
</tr>
</tbody>
</table>

**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
## 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>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaplastic (malignant) meningioma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic astrocytoma, IDH mutant</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic ependymoma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic ganglioglioma</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic oligodendroglioma IDH mutant and 1p/19q-codeleted</td>
<td>3</td>
</tr>
<tr>
<td>Anaplastic pleomorphic xanthroastrocytoma</td>
<td>3</td>
</tr>
<tr>
<td>Angiocentric glioma</td>
<td>1</td>
</tr>
<tr>
<td>Atypical choroid plexus papilloma</td>
<td>2</td>
</tr>
<tr>
<td>Atypical meningeoma</td>
<td>2</td>
</tr>
<tr>
<td>Atypical teratoid/rhabdoid tumor</td>
<td>4</td>
</tr>
<tr>
<td>Central neurocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Cerebellar liponeurocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Chordoid glioma of third ventricle</td>
<td>2</td>
</tr>
<tr>
<td>Choroid plexus carcinoma</td>
<td>3</td>
</tr>
<tr>
<td>Choroid plexus papilloma</td>
<td>1</td>
</tr>
<tr>
<td>CNS embryonal tumor NOS</td>
<td>4</td>
</tr>
<tr>
<td>CNS embryonal tumor with rhabdoid features</td>
<td>4</td>
</tr>
<tr>
<td>Craniopharyngioma</td>
<td>1</td>
</tr>
<tr>
<td>Desmoplastic infantile astrocytoma and ganglioglioma</td>
<td>1</td>
</tr>
<tr>
<td>Diffuse astrocytoma, IDH mutant</td>
<td>2</td>
</tr>
<tr>
<td>Diffuse midline glioma, H3K27M-mutant</td>
<td>4</td>
</tr>
<tr>
<td>Dysembryoplastic neuroepithelial tumor</td>
<td>1</td>
</tr>
<tr>
<td>Dysplastic gangliocytoma of cerebellum (D=Lhemite-Duclos)</td>
<td>1</td>
</tr>
<tr>
<td>Ependymoma</td>
<td>2</td>
</tr>
<tr>
<td>Ependymoma, RELA fusion-positive</td>
<td>2 or 3</td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Note:</strong> Tissue/pathology reports or CAP report will specify whether this is WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Extraventricular neurocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Gangliocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Glioblastoma, IDH mutant</td>
<td>4</td>
</tr>
<tr>
<td>Glioblastoma, IDH wildtype</td>
<td>4</td>
</tr>
<tr>
<td>Granular cell tumor</td>
<td>1</td>
</tr>
<tr>
<td>Hemangioblastoma</td>
<td>1</td>
</tr>
<tr>
<td>Malignant peripheral nerve sheath tumor (MPNST)</td>
<td>2, 3, or 4</td>
</tr>
<tr>
<td><strong>Note:</strong> Tissue/pathology reports or CAP report will specify whether this is WHO Grade 2, 3, or 4</td>
<td></td>
</tr>
<tr>
<td>Medulloblastoma (including all subtypes)</td>
<td>4</td>
</tr>
<tr>
<td>Medulloepithelioma</td>
<td>4</td>
</tr>
<tr>
<td>Meningioma</td>
<td>1</td>
</tr>
<tr>
<td>Myxopapillary ependymoma</td>
<td>1</td>
</tr>
<tr>
<td>Neurofibroma</td>
<td>1</td>
</tr>
<tr>
<td>Oligodendroglioma IDH mutant and 1p/19q-codeleted</td>
<td>2</td>
</tr>
<tr>
<td>Papillary glioneuronal tumor</td>
<td>1</td>
</tr>
<tr>
<td><strong>Note:</strong> Tissue/pathology report or CAP will specify whether it is WHO Grade 2 or 3</td>
<td></td>
</tr>
<tr>
<td>Papillary tumor of the pineal region</td>
<td>2 or 3</td>
</tr>
<tr>
<td>Perineuroma</td>
<td>1</td>
</tr>
<tr>
<td>Pilocytic astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td><strong>Note:</strong> ICD-O-3 lists a behavior code of /1. US and Canada collect as /3.</td>
<td></td>
</tr>
<tr>
<td>Pineal parenchymal tumor of intermediate differentiation</td>
<td>2 or 3</td>
</tr>
<tr>
<td><strong>Note:</strong> Tissue/pathology or CAP synoptic report will specify WHO Grade 2 or 3</td>
<td></td>
</tr>
</tbody>
</table>

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)  
Non-Malignant CNS Solid Tumor Rules 2018  
July 2019 Update
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>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pineoblastoma</td>
<td>4</td>
</tr>
<tr>
<td>Pineocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Pituicytoma</td>
<td>1</td>
</tr>
<tr>
<td>Pleomorphic xanthroastrocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Rosette-forming glioneuronal tumor</td>
<td>1</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>1</td>
</tr>
<tr>
<td>Solitary fibrous tumor/hemangiopericytoma</td>
<td>1, 2, or 3</td>
</tr>
</tbody>
</table>

*Note:* Tissue/pathology will specify WHO Grade 1, 2, or 3

<table>
<thead>
<tr>
<th>Histology</th>
<th>WHO Grade</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spindle cell oncocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Subependymal giant cell astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td>Subependymoma</td>
<td>1</td>
</tr>
</tbody>
</table>
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.
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)

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
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.
## Table 2: Reportable Primary Sites

**Column 1** lists the reportable primary site term.  
**Column 2** lists the site code for the reportable primary site.

<table>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
</table>
| **Brain**                   | Brain NOS C719  
Brain stem C717  
Cerebellum NOS C716  
Cerebrum C710  
Frontal lobe C711  
Occipital lobe C714  
Overlapping lesion of brain and central nervous system C718  
Parietal lobe C713  
Temporal lobe C712  
Ventricle NOS C715 |
| **Cranial Nerves**          | Abducent (cranial nerve VI) C725  
Accessory (cranial nerve XI) C725  
Acoustic (cranial nerve VIII) C724  
Crani al nerve NOS C725  
Facial (cranial nerve VII) C725  
Glossopharyngeal (cranial nerve IX) C725  
Hypoglossal (cranial nerve XII) C725  
Oculomotor (cranial nerve III) C725  
Olfactory (cranial nerve I C722)  
Optic (cranial nerve II) C723  
Trigeminal (cranial nerve V) C725  
Troc hlear (cranial nerve IV) C725  
Vagus (cranial nerve X) C725 |
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>
<thead>
<tr>
<th>Site Group</th>
<th>Reportable Subsite Terms and Code</th>
</tr>
</thead>
</table>
| Ill-Defined Sites Central Nervous System | Nervous system NOS C729  
Overlapping lesion of brain and central nervous system C728 |
| Intracranial Duct and Glands       | Craniopharyngeal duct C752  
Pineal gland C753  
Pituitary gland C751 |
| Meninges                           | Cerebral meninges C700  
Meninges NOS C709  
Spinal meninges C701 |
| Spinal Sites                        | Cauda equina/conus medullaris/filum terminale C721  
Meninges NOS C709  
Spinal cord C720  
Spinal meninges C701 |
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

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cranial nerve NOS C725</td>
<td></td>
<td><strong>Within cranium</strong>, unknown which nerve</td>
<td></td>
</tr>
<tr>
<td>Olfactory CN 1 C722</td>
<td>Cribriform plate</td>
<td>Surface of the <strong>brain</strong></td>
<td>Originates on the <strong>olfactory mucosa</strong> of the <strong>nasal cavity</strong>, then travels through the <strong>cribriform plate</strong> of the <strong>ethmoid bone</strong></td>
</tr>
<tr>
<td>Optic CN 2 C723</td>
<td>Optic canal</td>
<td><strong>Always reportable</strong>: CN2 is unique because it is intradural, covered with the meninges/dura and all portions are <strong>reportable</strong>.</td>
<td></td>
</tr>
<tr>
<td>Oculomotor CN 3 C725</td>
<td>Superior orbital fissure</td>
<td>Originates in the <strong>midbrain</strong>.</td>
<td>After exiting the superior orbital fissure, the nerve enters the <strong>orbit</strong>.</td>
</tr>
</tbody>
</table>
## Name and CN #

<table>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trochlear CN 4</td>
<td>Superior orbital fissure</td>
<td>Arises from the <em>dorsal brain stem</em>, loops around the brainstem and passes anteriorly within the <em>subarachnoid space</em>. It travels between the <em>superior cerebellar and posterior cerebral arteries</em> and through the <em>dura</em>, enters <em>cavernous sinus</em>.</td>
<td>Enters the <em>orbital fissure</em>.</td>
</tr>
<tr>
<td>C725</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trigeminal CN 5</td>
<td>The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina, the superior orbital fissure, the foramen rotundum and the foramen ovale.</td>
<td>CN5 originates in the <em>pons</em>. Upon leaving the pons it enters a <em>small fossa</em> posterior and inferolateral to the cavernous sinus called <em>Meckel's (trigeminal) cave</em>.</td>
<td><em>Ophthalmic nerve branch</em> crosses the <em>pterygopalatine fossa</em>, inclines laterally on the back of the <em>maxilla</em>, and enters the <em>orbit</em> through the inferior orbital fissure where it is called the <em>infraorbital nerve</em>. It ends beneath the <em>quadatus labii superius</em>, and divides into multiple branches that spread to the side of the <em>nose</em>, the lower <em>eyelid</em>, and the upper <em>lip</em>. <em>Maxillary nerve</em> leaves foramen rotundum and traverses the <em>infraorbital groove</em> and <em>canal</em> in the floor of the <em>orbit</em>, and appears on the face at the infraorbital foramen. <em>Mandibular nerve</em> leaves via the <em>foremen ovale</em> travels along the <em>mandibular groove</em>.</td>
</tr>
<tr>
<td>C725</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*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.
### 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>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abducent CN 6 C725</td>
<td>Cranial meninges</td>
<td>Exits brainstem at junction of <strong>pons</strong> and the <strong>medulla</strong>, enters the <strong>subarachnoid</strong> space and runs upward between the pons and the <strong>clivus</strong> entering the <strong>cavernous sinus</strong>.</td>
<td><strong>Dorello's canal</strong> and travels to the tip of the <strong>temporal bone</strong>. Enters <strong>orbit</strong></td>
</tr>
<tr>
<td>Facial CN 7 C725</td>
<td>Internal acoustic meatus</td>
<td><strong>CN7</strong> originates in the <strong>pons</strong>, along the posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity)</td>
<td>Enters the <strong>temples</strong> through the <strong>internal auditory meatus</strong> and runs through the <strong>facial canal</strong>.</td>
</tr>
<tr>
<td>Acoustic or vestibulocochlear CN 8 C724</td>
<td>Internal acoustic meatus</td>
<td>Originates in the <strong>brain stem (medulla oblongata) between</strong> the base of the brain (<strong>pons</strong>) and the <strong>spinal cord</strong>. Both the <strong>vestibular</strong> branch and the <strong>cochlear</strong> branch are located in the <strong>inner ear</strong>.</td>
<td></td>
</tr>
<tr>
<td>Glossopharyngeal CN 9 C725</td>
<td>Jugular foramen</td>
<td><strong>Originates</strong> in the anterior portion of the <strong>medulla oblongata</strong></td>
<td><strong>Jugular foramen</strong> Between the <strong>internal jugular vein</strong> and <strong>internal carotid artery</strong> Lies on the <strong>stylopharyngeus</strong> and <strong>middle pharyngeal constrictor muscle</strong> Passes under the <strong>hypoglossus muscle</strong> <strong>Palatine tonsil</strong> Extends to <strong>mucous glands</strong> of the <strong>mouth</strong>, and <strong>base of the tongue</strong></td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vagus CN 10 C725</td>
<td>Jugular foramen</td>
<td>The vagus nerve originates from the medulla of the brainstem.</td>
<td>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.</td>
</tr>
<tr>
<td>Accessory CN 11 C725</td>
<td>Jugular foramen</td>
<td>The nerve enters the foramen magnum or lateral aspect of the medulla oblongata.</td>
<td>The spinal accessory nerve originates in the neurons of the upper spinal cord, specifically C1-C5/C6 spinal nerve roots. The fibers of the spinal accessory nerve coalesce to form spinal rootlets, roots, and finally the spinal accessory nerve itself. The nerve exits the skull through the jugular foramen. It then runs along the internal carotid artery within the neck and reaches the sternocleidomastoid muscle and the trapezius.</td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Name and CN #</th>
<th>Exits Cranium Through</th>
<th>Reportable Portions of CN</th>
<th>Non-Reportable Portions of CN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoglossal CN12 C725</td>
<td>Hypoglossal canal</td>
<td>CN12 starts in the hypoglossal nucleus of the brainstem,</td>
<td>CN12 exits the hypoglossal canal, traveling between the carotid artery and jugular vein, ending under the tongue.</td>
</tr>
</tbody>
</table>

Jump to [Multiple Primary Rules](#)  
Jump to [Histology Coding Rules](#)
## 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).

<table>
<thead>
<tr>
<th>Non-reportable Histology Term</th>
<th>Non-reportable Histology Code</th>
<th>Definitions and Sites</th>
</tr>
</thead>
</table>
| Carcinomas                   | 8010-8060, 8071-8671, 8940-8941 | Brain C710-C719  
|                              |                              | **Site/histology edit** carcinomas/brain |
| Carcinomas                   | 8010-8671, 8940-8941          | Cerebral meninges, spinal meninges, meninges NOS C700-C709  
|                              |                              | **Site/histology edit** carcinomas/meninges |
| Carcinomas                   | 8010-8671, 8940-8941          | C721-C729 (Other central nervous system)  
|                              |                              | **Site/histology edit** carcinomas/other CNS |
| Colloid cyst                  | No code                      | **Site/histology edit** carcinomas/other CNS |
| Epidermoid tumor/cyst        | No code                      | **Site/histology edit** carcinomas/other CNS |
| 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                       | Occurs in hypothalamus |
| Hypothalamic hamartoma       | No code                      | Occurs in hypothalamus |
| 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 |
| 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 INTRACRANIALY.

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

<table>
<thead>
<tr>
<th>Histology Term and Code</th>
<th>Most Common Intracranial Primary Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angiocentric glioma 9431/1*</td>
<td>Cerebrum C710</td>
</tr>
<tr>
<td>Choroid plexus papilloma 9390/0</td>
<td>Intraventricular site (lateral/third ventricle C715 and IV ventricle C717)</td>
</tr>
<tr>
<td>(Capillary) hemangioblastoma 9161/1</td>
<td>Cerebellum C716, cerebrum (rare) C710</td>
</tr>
<tr>
<td>Craniopharyngioma 9350/1</td>
<td>Craniopharyngeal duct C752, pituitary gland, sella turcica C751</td>
</tr>
<tr>
<td>Dermoid cyst 9084/0</td>
<td>Pineal gland C753, suprasellar C719</td>
</tr>
<tr>
<td>Desmoplastic infantile astrocytoma and ganglioglioma 9412/1</td>
<td>Cerebrum/supratentorial brain NOS C710</td>
</tr>
<tr>
<td>Dysembryoplastic neuroepithelial tumor (DNT) 9413/0</td>
<td>Cerebrum C710, temporal lobe C712</td>
</tr>
<tr>
<td>Dysplastic gangliocytoma 9493/0</td>
<td>Cerebellum C716</td>
</tr>
<tr>
<td>Meningioma (rare) 9530/0</td>
<td>Intraventricular C715</td>
</tr>
<tr>
<td>Myxopapillary ependymoma 9394/1</td>
<td>4th ventricle C717</td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>Histology Term and Code</th>
<th>Most Common Intracranial Primary Site</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1</td>
<td>Posterior fossa C719, cerebrum C710</td>
</tr>
<tr>
<td>Pineocytoma 9361/1</td>
<td>Pineal gland C753</td>
</tr>
<tr>
<td>Pituicytoma 9432/1*</td>
<td>Pituitary gland C751, sella turcica C751, suprasellar C719</td>
</tr>
<tr>
<td>Pituitary adenoma 8272/0</td>
<td>Pituitary gland C751</td>
</tr>
<tr>
<td>Prolactinoma 8271/0</td>
<td>Pituitary gland C751</td>
</tr>
<tr>
<td>Subependymal giant cell tumor (SEGA) 9384/1</td>
<td>Lateral ventricles C715</td>
</tr>
<tr>
<td>Subependymoma 9383/1</td>
<td>Intraventricular site (lateral/third ventricle, rare C715 and IV ventricle C717)</td>
</tr>
</tbody>
</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 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.

<table>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
</table>
| Angiocentric glioma 9431/1*          | Angiocentric neuroepithelial tumor  
Monomorphic angiocentric glioma       |                                          |
| Benign fibrous histiocyteoma 8830/0  |                                      |
| Central neurocytoma 9506/1           | Cerebellar liponeurocytoma  
Extraventricular neurocytoma          |                                          |
| Chondroma 9220/0                     |                                      |
| Chordoid glioma of the third ventricle 9444/1 |               |
| Choroid plexus papilloma 9390/0      | Atypical choroid plexus papilloma 9390/1 |
| Craniopharyngioma 9350/1             | Adamantinomatous craniopharyngioma 9351/1  
Papillary craniopharyngioma 9352/1    |                                          |
## 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>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Desmoplastic infantile astrocytoma and ganglioglioma 9412/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dysembryoplastic neuroepithelial tumor 9413/0</td>
<td>DNT</td>
<td></td>
</tr>
<tr>
<td>Gangliocytoma 9492/0</td>
<td></td>
<td>Dysplastic cerebellar gangliocytoma/Lhermitte-Duclos disease 9493/0</td>
</tr>
<tr>
<td>Ganglioglioma 9505/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Granular cell tumor of the sellar region 9582/0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hemangioblastoma 9161/1</td>
<td></td>
<td>Capillary hemangioblastoma</td>
</tr>
<tr>
<td>Hemangioma 9120/0</td>
<td></td>
<td>Cavernous hemangioma 9121/0</td>
</tr>
<tr>
<td>Leiomyoma 8890/0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lipoma 8850/0</td>
<td></td>
<td>Hibernoma 8880/0</td>
</tr>
<tr>
<td>Meningeal melanocytosis 8728/0</td>
<td></td>
<td>Meningeal melanocytoma 8728/1</td>
</tr>
<tr>
<td>Meningioma 9530/0</td>
<td>Lymphoplasmacyte-rich meningioma Metaplastic meningioma Microcystic meningioma Secretory meningioma</td>
<td>Angiomatous meningioma 9534/0 Atypical meningioma 9539/1 Clear cell/chordoid meningioma 9538/1 Fibrous meningioma 9532/0 Meningothelial meningioma 9531/0 Psammomatous meningioma 9533/0 Transitional meningioma 9537/0</td>
</tr>
<tr>
<td>Myofibroblastoma 8825/0</td>
<td></td>
<td>Inflammatory myofibroblastic tumor 8825/1</td>
</tr>
<tr>
<td>Myxopapillary ependymoma 9394/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurofibroma 9540/0</td>
<td>Atypical neurofibroma</td>
<td>Plexiform neurofibroma 9550/0</td>
</tr>
<tr>
<td>Optic glioma/pilocytic astrocytoma 9421/1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Osteoma 9180/0</td>
<td></td>
<td></td>
</tr>
</tbody>
</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>
<thead>
<tr>
<th>NOS/Specific Histology Term and Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants Histology Term and Codes</th>
</tr>
</thead>
</table>
| Papillary glioneuronal tumor 9509/1* | Diffuse leptomeningeal glioneuronal tumor  
Rosette-forming glioneuronal tumor | | |
| Paraganglioma 8693/1 | | |
| Perineurioma 9571/0 | | |
| Pineocytoma 9361/1 | | |
| Pituicytoma 9432/1* | | |
| Pituitary adenoma 8272/0 | Corticotroph | |
| Prolactinoma 8271/0 | | |
| Rhabdomyoma 8900/0 | | |
| Schwannoma 9560/0 | Cellular schwannoma  
Neurilemoma  
Neurinoma  
Plexiform schwannoma | Melanotic schwannoma 9560/1* |
| Solitary fibrous tumor Grade 1 8815/0 | Hemangiopericytoma Grade 1 | Solitary fibrous tumor/hemangiopericytoma  
Grade 2 8815/1* |
| Spindle cell oncocytoma 8290/0 | | |
| Subependymal giant cell astrocytoma 9384/1 | | |
| Subependymoma 9383/1 | | |
| Teratoma 9080/1 | | |
Conflicting Information on Pathology Report(s)

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

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

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

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

- **Single pathology report:**
  - **Multiple pathologists** within the institution review the slides
  - Slides are sent for **outside review** and the information from the **consulting** lab **conflicts** with the **original** pathology report

- **Multiple pathology reports:** The **first report** is from a biopsy and the **second report** is from a resection. Code the histology and/or behavior from the resection.
Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Table 7: Paired Sites

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

<table>
<thead>
<tr>
<th>Paired Sites and Codes</th>
<th>Codes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acoustic nerve</td>
<td>C724</td>
</tr>
<tr>
<td>Cerebral meninges</td>
<td>C700</td>
</tr>
<tr>
<td>Cerebrum</td>
<td>C710</td>
</tr>
<tr>
<td>Cranial nerves</td>
<td>C725</td>
</tr>
<tr>
<td>Frontal lobe</td>
<td>C711</td>
</tr>
<tr>
<td>Occipital lobe</td>
<td>C714</td>
</tr>
<tr>
<td>Olfactory nerve</td>
<td>C722</td>
</tr>
<tr>
<td>Optic nerve</td>
<td>C723</td>
</tr>
<tr>
<td>Parietal lobe</td>
<td>C713</td>
</tr>
<tr>
<td>Temporal lobe</td>
<td>C712</td>
</tr>
</tbody>
</table>

**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.
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.

<table>
<thead>
<tr>
<th>Original Histology and Code</th>
<th>Transformed Histology and Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chondroma 9220/0</td>
<td>Chondrosarcoma 9220/3</td>
</tr>
<tr>
<td>Ganglioglioma 9505/1</td>
<td>Anaplastic ganglioglioma 9505/3</td>
</tr>
<tr>
<td>Hemangioma 9120/0</td>
<td>Angiosarcoma 9120/3</td>
</tr>
<tr>
<td>Hemangiopericytoma 9150/1</td>
<td>Anaplastic hemangiopericytoma 9150/3</td>
</tr>
<tr>
<td>Leiomyoma 8890/0</td>
<td>Leiomyosarcoma 8890/3</td>
</tr>
<tr>
<td>Lipoma 8850/0</td>
<td>Liposarcoma 8850/3</td>
</tr>
<tr>
<td>Osteoma 9180/0</td>
<td>Osteosarcoma 9180/3</td>
</tr>
<tr>
<td>Perineurioma 9571/0</td>
<td>Malignant perineurioma 9571/3</td>
</tr>
<tr>
<td>Rhabdomyoma 8900/0</td>
<td>Rhabdomyosarcoma 8900/3</td>
</tr>
<tr>
<td>Teratoma 9080/1</td>
<td>Immature teratoma 9080/3</td>
</tr>
<tr>
<td>Teratoma, mature 9080/0</td>
<td>Immature teratoma 9080/3</td>
</tr>
</tbody>
</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)
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)

A.D.A.M illustration used with licensed permission. All rights reserved.
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 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** 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.

\(^1\) Prepare one abstract. Use the histology rules to assign the appropriate histology code.
### 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 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 (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.
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)

- 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 abstract 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\(^i\) 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\(^1\) 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).
Rule M7  Abstract multiple primaries 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 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 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** 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** 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 8860/0 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\(^i\) when separate/non-contiguous tumors are on different rows in Table 6 in the Equivalent Terms and Definitions. Timing is irrelevant.  
\textit{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.  
\textit{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

---

\(^i\) 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.

\(^i\) 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**

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

2. Code the histology assigned by the physician. 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.
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 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 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/documentated
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)

**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

<table>
<thead>
<tr>
<th>Apparently</th>
<th>Most likely</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appears</td>
<td>Presumed</td>
</tr>
<tr>
<td>Comparable with</td>
<td>Probable</td>
</tr>
<tr>
<td>Compatible with</td>
<td>Suspect(ed)</td>
</tr>
<tr>
<td>Consistent with</td>
<td>Suspicious (for)</td>
</tr>
<tr>
<td>Favor(s)</td>
<td>Typical (of)</td>
</tr>
<tr>
<td>Malignant appearing</td>
<td></td>
</tr>
</tbody>
</table>

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

Jump to [Equivalent Terms and Definitions](#)
Jump to [Multiple Primary Rules](#)
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)

Jump to Equivalent Terms and Definitions
Jump to Multiple Primary Rules

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 8860/0 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.
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)

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 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.
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 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 8860/0 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)

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.
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.
### 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.

Priority for Coding Primary Site

The following list is 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 discontinuous 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)
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)

4. Code Urinary System NOS C689 when there are multiple discontinuous 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>
<thead>
<tr>
<th>Site Term and code</th>
<th>Synonyms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bladder, anterior wall C673</td>
<td>-</td>
</tr>
<tr>
<td>Bladder, dome C671</td>
<td>Roof</td>
</tr>
<tr>
<td></td>
<td>Vault</td>
</tr>
<tr>
<td></td>
<td>Vertex</td>
</tr>
<tr>
<td>Bladder, lateral wall C672</td>
<td>Lateral to ureteral orifice</td>
</tr>
<tr>
<td></td>
<td>Left wall</td>
</tr>
<tr>
<td></td>
<td>Right wall</td>
</tr>
<tr>
<td></td>
<td>Sidewall</td>
</tr>
<tr>
<td>Bladder neck C675</td>
<td>Internal urethral orifice</td>
</tr>
<tr>
<td></td>
<td>Vesical neck</td>
</tr>
<tr>
<td>Bladder NOS C679</td>
<td>Lateral posterior wall (no hyphen)</td>
</tr>
<tr>
<td>Bladder, overlapping lesion C678</td>
<td>Fundus</td>
</tr>
<tr>
<td></td>
<td>Lateral-posterior wall (hyphen)</td>
</tr>
<tr>
<td>Bladder, posterior wall C674</td>
<td>-</td>
</tr>
</tbody>
</table>
### 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>
<thead>
<tr>
<th>Site Term and code</th>
<th>Synonyms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bladder, trigone C670</td>
<td>Base of bladder</td>
</tr>
<tr>
<td></td>
<td>Below interureteric crest</td>
</tr>
<tr>
<td></td>
<td>Below interureteric field</td>
</tr>
<tr>
<td></td>
<td>Below interureteric ridge</td>
</tr>
<tr>
<td></td>
<td>Floor of bladder</td>
</tr>
<tr>
<td>Bladder, urachus C677</td>
<td>Mid umbilical ligament</td>
</tr>
<tr>
<td>Bladder, ureteric orifice C676</td>
<td>Just above ureteric orifice</td>
</tr>
<tr>
<td>Overlapping lesion of urinary organs C688</td>
<td>-</td>
</tr>
<tr>
<td>Paraurethral gland C681</td>
<td>-</td>
</tr>
<tr>
<td>Renal pelvis C659</td>
<td>Pelvis of kidney</td>
</tr>
<tr>
<td></td>
<td>Pelviureteric junction</td>
</tr>
<tr>
<td></td>
<td>Renal calyces</td>
</tr>
<tr>
<td></td>
<td>Renal calyx</td>
</tr>
<tr>
<td>Ureter C669</td>
<td>-</td>
</tr>
<tr>
<td>Urethra C680</td>
<td>Cowper gland</td>
</tr>
<tr>
<td></td>
<td>Prostatic utricle</td>
</tr>
<tr>
<td></td>
<td>Urethral gland</td>
</tr>
<tr>
<td>Urinary system NOS C689</td>
<td>-</td>
</tr>
</tbody>
</table>
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 using the Solid Tumor Rules, rhabdomyosarcoma and embryonal rhabdomyosarcoma are treated the same as all NOS and subtypes/variants.

**Table begins on next page**
### Specific and NOS Histology Codes

<table>
<thead>
<tr>
<th>Code</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Adenocarcinoma NOS 8140 | Mixed adenocarcinoma  
Urachal adenocarcinoma | Clear cell carcinoma 8310  
Endometrioid carcinoma 8380  
Enteric adenocarcinoma 8144  
Mucinous adenocarcinoma 8480 |
| Malignant melanoma 8720/3 |                                      |                                                       |
| Malignant perivascular epithelioid cell tumor 8714/3 | Malignant PEComa |                                                       |
| Sarcoma NOS 8800/3 |                                      | Angiosarcoma 9120/3  
Chondrosarcoma 9220/3  
Leiomyosarcoma 8890/3  
Liposarcoma 8850/3  
Malignant peripheral nerve sheath tumor (MPNST) 9540/3  
Pleomorphic sarcoma 8802/3  
Rhabdomyosarcoma 8900/3  
Embryonal rhabdomyosarcoma/sarcoma botryoides 8910/3 |                                                       |
| Small cell neuroendocrine carcinoma 8041 | Neuroendocrine carcinoma  
SmCC | Large cell neuroendocrine tumor 8013  
Well-differentiated neuroendocrine tumor 8240 |
| Squamous cell carcinoma 8070 | Pure squamous cell carcinoma  
SCC | Verrucous carcinoma 8051 |

Table continues on next page
<table>
<thead>
<tr>
<th>Specific and NOS Histology Codes</th>
<th>Synonyms</th>
<th>Subtypes/Variants</th>
</tr>
</thead>
</table>
| Urothelial carcinoma 8120        | Clear cell (glycogen-rich) urothelial carcinoma 8120/3  
                             Infiltrating urothelial carcinoma 8120/3  
                             Infiltrating urothelial carcinoma with divergent differentiation 8120/3  
                             Infiltrating urothelial carcinoma with endodermal sinus lines 8120/3  
                             Infiltrating urothelial carcinoma with glandular differentiation 8120/3  
                             Infiltrating urothelial carcinoma with squamous differentiation 8120/3  
                             Infiltrating urothelial carcinoma with trophoblastic differentiation 8120/3  
                             Lipid-rich urothelial carcinoma 8120/3  
                             Microcystic urothelial carcinoma 8120/3  
                             Nested urothelial carcinoma 8120/3  
                             Plasmacytoid urothelial carcinoma 8120/3  
                             Urothelial carcinoma in situ 8120/2 | Giant cell urothelial carcinoma 8031/3  
                             Lymphoepithelioma-like urothelial carcinoma 8082/3  
                             Plasmacytoid/signet ring cell/diffuse variant  
                             Papillary urothelial (transitional cell) carcinoma  
                             in situ 8130/2  
                             invasive 8130/3  
                             Micropapillary urothelial carcinoma 8131/3  
                             Poorly differentiated carcinoma 8020/3  
                             Sarcomatoid urothelial carcinoma 8122/3 |

**Note 1:** Previously called **transitional cell** carcinoma, a term that is no longer recommended.

**Note 2:** Micropapillary 8131 is a subtype/variant of papillary urothelial carcinoma 8130. It is an invasive /3 neoplasm with aggressive behavior.
### 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.

<table>
<thead>
<tr>
<th>Histology Term and Code</th>
<th>Synonyms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Benign perivascular epithelioid cell tumor <strong>8714/0</strong></td>
<td>Benign PEComa</td>
</tr>
<tr>
<td>Granular cell tumor <strong>9580/0</strong></td>
<td></td>
</tr>
<tr>
<td>Hemangioma <strong>9120/0</strong></td>
<td></td>
</tr>
<tr>
<td>Inflammatory myofibroblastic tumor <strong>8825/1</strong></td>
<td></td>
</tr>
<tr>
<td>Inverted urothelial papilloma <strong>8121/0</strong></td>
<td></td>
</tr>
<tr>
<td>Leiomyoma <strong>8890/0</strong></td>
<td></td>
</tr>
<tr>
<td>Melanosis <strong>No code</strong></td>
<td></td>
</tr>
<tr>
<td>Neurofibroma <strong>9540/0</strong></td>
<td></td>
</tr>
<tr>
<td>Nevus <strong>8720/0</strong></td>
<td></td>
</tr>
<tr>
<td>Papillary urothelial neoplasm of low-malignant potential <strong>8130/1</strong></td>
<td></td>
</tr>
<tr>
<td>Paraganglioma <strong>8693/1</strong></td>
<td>Extra-adrenal pheochromocytoma</td>
</tr>
<tr>
<td>Solitary fibrous tumor <strong>8815/1</strong></td>
<td></td>
</tr>
<tr>
<td>Squamous cell papilloma <strong>8052/0</strong></td>
<td>Keratotic papilloma</td>
</tr>
<tr>
<td>Urothelial dysplasia <strong>No code</strong></td>
<td></td>
</tr>
<tr>
<td>Urothelial papilloma <strong>8120/0</strong></td>
<td></td>
</tr>
<tr>
<td>Villous adenoma <strong>8261/0</strong></td>
<td></td>
</tr>
</tbody>
</table>
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)

Illustrations

A.D.A.M. illustration used with licensed permission. All rights reserved.
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)

www.MedicineNet.com
Illustration used with licensed permission. All rights reserved.
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)

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

Bladder Tumor

Bladder Wall

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

Microscopic Structure of the Ureter

- Lumen
- Adventitia
- Circular layer
- Longitudinal layer
- Transitional epithelium
- Lamina propria
- Mucosa
- Muscularis

Jump to Multiple Primary Rules
Jump to Histology Coding Rules
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 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\(^1\) 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.

\(^1\)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\(^2\) 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\(^d\) 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\(^d\) 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\(^d\) 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
Rule M7  Abstract a **single primary** 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** 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** when the patient has multiple **invasive** urothelial cell carcinomas in the **bladder**. All tumors are **either**:

- Multiple occurrences of urothelial or urothelial subtypes (with exception of micropapillary) **OR**
- Multiple occurrences of micropapillary

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

*Note 2:* Abstract only one \( /3 \) invasive urothelial bladder primary **AND** only one micropapillary urothelial 8131/3 bladder primary per the patient’s lifetime.

- An occurrence of micropapillary and an occurrence of urothelial carcinoma would be multiple primaries (see previous rules).
Renal Pelvis, Ureter, Bladder, and Other Urinary Multiple Primary Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M10  Abstract multiple primaries\(^{d}\) 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\(^{d}\) 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\(^\text{ii}\) 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\(^\text{ii}\) 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\(^\text{ii}\) when the ICD-O site code differs at the second (C\(\text{X}\)xx) and/or third (C\(\text{x}\)Xx) character.

Rule M15  
Abstract a single primary\(^\text{i}\) 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)

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\(^1\) (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\(^1\) (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\(^1\) 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.

\(^1\)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

   Note 1: Histology changes do occur following immunotherapy, chemotherapy and radiation therapy.
   Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

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 **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/documentated

      **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.**
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)

List of Ambiguous Terminology

- Apparently
- Appears
- Comparable with
- Compatible with
- Consistent with
- Favor(s)
- Malignant appearing

- Most likely
- Presumed
- Probable
- Suspect(ed)
- Suspicious (for)
- Typical (of)

4. **DO NOT CODE** histology when described as:

- Architecture
- Configuration
- Foci; focus; focal
- 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 tumor**.

**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/varieties of small cell neuroendocrine carcinoma
- Subtype/variant of small cell neuroendocrine mixed with any other carcinoma (does not apply to sarcoma)

*Example:* Diagnosis from TURB is urothelial carcinoma and small cell neuroendocrine carcinoma. Code mixed small cell carcinoma 8045.
Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

**Rule H5** Code mixed urothelial carcinoma as follows:
- **Code 8120** when urothelial is mixed with:
  - Adenocarcinoma or adenocarcinoma subtypes
  - Squamous cell carcinoma or squamous cell carcinoma subtypes
- **Code 8130** when papillary urothelial is mixed with:
  - Adenocarcinoma or adenocarcinoma subtypes
  - Squamous cell carcinoma or squamous cell carcinoma subtypes
- **Code 8131/3** when micropapillary urothelial is mixed with:
  - Adenocarcinoma or adenocarcinoma subtypes
  - Squamous cell carcinoma or squamous cell carcinoma subtypes

*Note:* Adenocarcinoma and subtypes/variants as well as squamous cell carcinoma and subtypes/variants are coded ONLY when pure (not mixed with any other histology).

*Example:* Pathology says majority of tumor is squamous cell carcinoma 8070/3 with a minority composed of papillary urothelial cell carcinoma 8130/3. Code the papillary urothelial cell carcinoma 8130/3. The squamous cell carcinoma is not pure and cannot be coded.

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

Code the histology using the rule that fits the case.
Renal Pelvis, Ureter, Bladder, and Other Urinary Histology Rules
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – 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.
The following sections are to be used for Other Sites and Cutaneous Melanoma cases diagnosed 2007-2020.

The 2007 General Instructions are to be used in conjunction with the Other Sites and Cutaneous Melanoma sections. DO NOT USE the Solid Tumor 2018 General Instructions for these cases.
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 12/31/2018

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

Jump to Multiple Primary Rules
Jump to Histology Rules
### 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.

<table>
<thead>
<tr>
<th>Site Code</th>
<th>Site or Subsite</th>
</tr>
</thead>
<tbody>
<tr>
<td>C384</td>
<td>Pleura</td>
</tr>
<tr>
<td>C400</td>
<td>Long bones of upper limb, scapula, and associated joints</td>
</tr>
<tr>
<td>C401</td>
<td>Short bones of upper limb and associated joints</td>
</tr>
<tr>
<td>C402</td>
<td>Long bones of lower limb and associated joints</td>
</tr>
<tr>
<td>C403</td>
<td>Short bones of lower limb and associated joints</td>
</tr>
<tr>
<td>C413</td>
<td>Rib, clavicle (excluding sternum)</td>
</tr>
<tr>
<td>C414</td>
<td>Pelvic bones (excluding sacrum, coccyx, symphysis pubis)</td>
</tr>
<tr>
<td>C441</td>
<td>Skin of the eyelid</td>
</tr>
<tr>
<td>C442</td>
<td>Skin of the external ear</td>
</tr>
<tr>
<td>C443</td>
<td>Skin of other and unspecified parts of the face (if midline, assign code 9)</td>
</tr>
<tr>
<td>C445</td>
<td>Skin of the trunk (if midline, assign code 9)</td>
</tr>
<tr>
<td>C446</td>
<td>Skin of upper limb and shoulder</td>
</tr>
<tr>
<td>C447</td>
<td>Skin of the lower limb and hip</td>
</tr>
<tr>
<td>C471</td>
<td>Peripheral nerves and autonomic nervous system of upper limb and shoulder</td>
</tr>
<tr>
<td>C472</td>
<td>Peripheral nerves and autonomic nervous system of the lower limb and hip</td>
</tr>
<tr>
<td>C491</td>
<td>Connective, subcutaneous, and other soft tissues of upper limb and shoulder</td>
</tr>
<tr>
<td>C492</td>
<td>Connective, subcutaneous, and other soft tissues of the lower limb and hip</td>
</tr>
<tr>
<td>C569</td>
<td>Ovary</td>
</tr>
<tr>
<td>C570</td>
<td>Fallopian tube</td>
</tr>
<tr>
<td>C620-C629</td>
<td>Testis</td>
</tr>
<tr>
<td>C630</td>
<td>Epididymis</td>
</tr>
<tr>
<td>C631</td>
<td>Spermatic cord</td>
</tr>
<tr>
<td>C690-C699</td>
<td>Eye and adnexa</td>
</tr>
<tr>
<td>C740-C749</td>
<td>Adrenal gland</td>
</tr>
<tr>
<td>C754</td>
<td>Carotid body</td>
</tr>
</tbody>
</table>
### 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.

<table>
<thead>
<tr>
<th>Column 1: Required Histology</th>
<th>Column 2: Combined with Histology</th>
<th>Column 3: Combination Term</th>
<th>Column 4: Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small cell carcinoma</td>
<td>Large cell carcinoma</td>
<td>Combined small cell carcinoma</td>
<td>8045</td>
</tr>
<tr>
<td></td>
<td>Adenocarcinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Squamous cell carcinoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Squamous carcinoma</td>
<td>Basal cell carcinoma</td>
<td>Basosquamous carcinoma</td>
<td>8094</td>
</tr>
<tr>
<td>Islet cell</td>
<td>Exocrine</td>
<td>Mixed islet cell and exocrine adenocarcinoma (pancreas)</td>
<td>8154</td>
</tr>
<tr>
<td>Acinar</td>
<td>Endocrine</td>
<td>Mixed islet cell and exocrine adenocarcinoma (pancreas)</td>
<td>8154</td>
</tr>
<tr>
<td>Hepatocellular carcinoma</td>
<td>Cholangiocarcinoma</td>
<td>Combined hepatocellular carcinoma and cholangiocarcinoma</td>
<td>8180</td>
</tr>
<tr>
<td>Adenocarcinoma</td>
<td>Carcinoid</td>
<td>Composite carcinoid</td>
<td>8244</td>
</tr>
<tr>
<td>Adenocarcinoma and two or more of the histologies from column 2 OR two or more of the histologies from column 2</td>
<td>Papillary Clear cell Mucinous (colloid) Signet ring Acinar</td>
<td>Adenocarcinoma with mixed subtypes Adenocarcinoma combined with other types of carcinoma</td>
<td>8255</td>
</tr>
</tbody>
</table>
### 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>
<thead>
<tr>
<th>Column 1: Required Histology</th>
<th>Column 2: Combined with Histology</th>
<th>Column 3: Combination Term</th>
<th>Column 4: Code</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gyn malignancies with two or more of the histologies in column 2</td>
<td>Clear cell Endometrioid Mucinous Papillary Serous Squamous Transitional (Brenner)</td>
<td>Mixed cell adenocarcinoma</td>
<td>8323</td>
</tr>
<tr>
<td>Papillary and Follicular</td>
<td></td>
<td>Papillary carcinoma, follicular variant</td>
<td>8340</td>
</tr>
<tr>
<td>Medullary</td>
<td>Follicular</td>
<td>Mixed medullary-follicular carcinoma</td>
<td>8346</td>
</tr>
<tr>
<td>Medullary</td>
<td>Papillary</td>
<td>Mixed medullary-papillary carcinoma</td>
<td>8347</td>
</tr>
<tr>
<td>Squamous carcinoma and Adenocarcinoma</td>
<td></td>
<td>Adenosquamous carcinoma</td>
<td>8560</td>
</tr>
<tr>
<td>Any combination of histologies in Column 2</td>
<td>Myxoid Round cell Pleomorphic</td>
<td>Mixed liposarcoma</td>
<td>8855</td>
</tr>
<tr>
<td>Embryonal rhabdomyosarcoma</td>
<td>Alveolar rhabdomyosarcoma</td>
<td>Mixed type rhabdomyosarcoma</td>
<td>8902</td>
</tr>
<tr>
<td>Teratoma</td>
<td>Embryonal carcinoma</td>
<td>Teratocarcinoma</td>
<td>9081</td>
</tr>
<tr>
<td>Teratoma and one or more of the histologies in Column 2</td>
<td>Seminoma Yolk sac tumor</td>
<td>Mixed germ cell tumor</td>
<td>9085</td>
</tr>
<tr>
<td>Choriocarcinoma</td>
<td>Teratoma Seminoma Embryonal</td>
<td>Choriocarcinoma combined with other germ cell elements</td>
<td>9101</td>
</tr>
</tbody>
</table>
Previous to 2007, tumors in sites on the same row were abstracted as a single primary.

<table>
<thead>
<tr>
<th>Code</th>
<th>Site Groupings</th>
</tr>
</thead>
<tbody>
<tr>
<td>C23</td>
<td>Gallbladder</td>
</tr>
<tr>
<td>C24</td>
<td>Other and unspecified parts of the biliary tract</td>
</tr>
<tr>
<td>C37</td>
<td>Thymus</td>
</tr>
<tr>
<td>C380</td>
<td>Heart</td>
</tr>
<tr>
<td>C381-3</td>
<td>Mediastinum</td>
</tr>
<tr>
<td>C388</td>
<td>Overlapping lesion of heart, mediastinum, and pleura</td>
</tr>
<tr>
<td>C51</td>
<td>Vulva</td>
</tr>
<tr>
<td>C52</td>
<td>Vagina</td>
</tr>
<tr>
<td>C577</td>
<td>Other specified female genital organs</td>
</tr>
<tr>
<td>C578-9</td>
<td>Unspecified female genital organs</td>
</tr>
<tr>
<td>C569</td>
<td>Ovary</td>
</tr>
<tr>
<td>C570</td>
<td>Fallopian tube</td>
</tr>
<tr>
<td>C571</td>
<td>Broad ligament</td>
</tr>
<tr>
<td>C572</td>
<td>Round ligament</td>
</tr>
<tr>
<td>C573</td>
<td>Parametrium</td>
</tr>
<tr>
<td>C574</td>
<td>Uterine adnexa</td>
</tr>
<tr>
<td>C60</td>
<td>Penis</td>
</tr>
<tr>
<td>C63</td>
<td>Other and unspecified male genital organs</td>
</tr>
<tr>
<td>C74</td>
<td>Adrenal gland</td>
</tr>
<tr>
<td>C75</td>
<td>Other endocrine glands and related structures</td>
</tr>
</tbody>
</table>
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 12/31/2018

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 (Cxxx) and/or third characters (Cxx) are multiple primaries. **
Example 1: A tumor in the penis C609 and a tumor in the rectum C209 have different second characters in their ICD-O-3 topography codes, so they are multiple primaries.
Example 2: A tumor in the cervix C539 and a tumor in the vulva C519 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) 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.

Jump to Equivalent Terms and Definitions
Jump to Histology Rules

Other Sites Solid Tumor Rules 2018
Updated 9/11/2018
For cases diagnosed 2007 and later
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 (xxxx), second (xxx) or third (xx) 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, Lymphomas and Leukemia

For cases diagnosed 1/1/2007 to 12/31/2018

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 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

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

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

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 history 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 *adeno*matous *polyp*), 8261 (adenocarcinoma in *villo*us *adenoma*), or 8263 (adenocarcinoma in *tubulo*villo*us* *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.


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

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

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

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.
Cutaneous melanoma starts in the melanocyte cells 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 go on to 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 melanomas**: are lumpy and often blue-black in color and may grow faster and spread downwards

These types account for the majority of melanomas occurring in the US population. For a more complete listing of histologic types of melanoma, see the *AJCC Cancer Staging Manual*, 6th Ed.

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.
Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2007 and later

- Type, subtype, predominantly, with features of, major, or with ____differentiation.
- Giant pigmented nevus, giant congenital nevus
- Mole, Nevus
- Mixed epithelioid and spindle cell melanoma (8770): Epithelioid melanoma and spindle cell melanoma

### 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
- Stage 0
- Tis

### Synonyms for Hutchinson freckle

- Circumscribed precancerous melanosis
- Intraepidermal malignant melanoma
- Lentigo maligna
- Precancerous melanosis of Dubreuilh
Amelanotic melanoma: A non-pigmented malignant melanoma.

Atypical melanocytic hyperplasia (dysplasia): Tumor-like lesion or condition may represent precursor stage or stage in development of melanoma. Not reportable.

Different lateralities: The right side of the body, the left side of the body and the midline are separate lateralities in the melanoma coding rules.

Evolving melanoma (borderline evolving melanoma): Evolving melanoma are tumors of uncertain biologic behavior. Histological changes of borderline evolving melanoma are too subtle for a definitive diagnosis of melanoma in situ. The tumors may be described as "proliferation of atypical melanocytes confined to epidermal and adnexal epithelium," "atypical intraepidermal melanocytic proliferation," "atypical intraepidermal melanocytic hyperplasia"; or “severe melanocytic dysplasia.” Not reportable.

Familial Atypical Multiple Mole Melanoma Syndrome (FAMM, FAM-M): An inherited condition identified when:
- Melanoma has been diagnosed in a family member, including grandparents, aunts, uncles, and cousins
- Several family members have large numbers of moles (often more than 50) which may be abnormal or atypical moles.

Giant pigmented nevus: Diameter larger than 20 cm; frequently covers large areas of the body in a garment-like fashion. The trunk, head and neck are the most common sites.

Junctional nevus: Smooth, hairless, light to dark brown mole. Can be slightly elevated, usually multiple and can occur on any part of the body. Melanocytes are confined to the dermo-epidermal junction.

Hypodermis: A subcutaneous layer of loose connective tissue containing a varying number of fat cells. Synonyms: subcutaneous fat; subcutis.

In-transit metastasis: Metastasis found in the lymphatic channels more than 2cm away from the primary melanoma, but not reaching the regional lymph nodes.
Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2007 and later

**Invasive tumor:** A tumor that penetrates the basement membrane and invades the dermis.

**Laterality:** For skin sites, laterality divides the body into a right and left half as though a line were drawn from mid forehead to mid pelvis and from mid skull to mid buttocks. A midline laterality describes a tumor that is in the center of the “line” drawn from the mid forehead to mid pelvis or from the mid skull to the mid buttocks; it is impossible to categorize the tumor as being on the right or left side of the body.

**Lentigo maligna:** Is a specific histologic type of in situ melanoma. It appears as a brown or black mottled, irregular, lesion with increased numbers of scattered atypical melanocytes in the epidermis. It usually occurs on the face.

**Lentigo maligna melanoma:** Is an invasive melanoma that begins as lentigo maligna, but usually after many years the dermis is invaded by the tumor. Once invasion has occurred, the lesion is called lentigo maligna melanoma.

**Midline:** The middle dividing line that separates the body into right and left sides.

**Most invasive:** The histology that has the greatest extension into the dermis or subcutaneous fat.

**Non-invasive tumor:** A tumor confined to epithelium (intraepithelial), in situ tumor, with no penetration below the basement membrane.

**Precancerous melanosis:** An obsolete term for lentigo maligna.

**Proliferation of atypical melanocytes confined to epidermis:** Number of (proliferation) pigmented cells (melanocytes) not showing the normal cell structure (atypical). Not reportable.

**Regressing melanoma:** The term “regressing melanoma” does not refer to a specific histology; it refers to the physical appearance and size of the lesion. A regressing melanoma is reacting to the body’s immune system by shrinking in size. Partial spontaneous regression is not an uncommon finding in invasive primary melanoma; partial regression can be an indicator of poor prognosis. Proven complete regression is very rare; one website stated that only 33 cases of total regression have been reported. A
regressive melanoma is usually thinner than it was originally. Although regression is a prognostic factor, the histologic type is more important for histology coding purposes. See Histology coding rules, Rule H5.

Satellite lesion or metastasis: Grossly evident metastatic skin lesion within the immediate vicinity (usually within 2 cm) of a primary malignant tumor; e.g., skin adjacent to primary malignant melanoma. This is a metastasis, not a separate primary.

Severe melanotic dysplasia: Tumor-like lesion or condition. Not reportable.

Skin Layers:
- Epidermis – upper surface, thin layer (outermost layer)
- Dermis – lower, intermediate thicker layer (intermediate layer)
- Hypodermis – also called subcutis or subcutaneous fat – lowest layer (innermost layer)
Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2007 and later

Illustrations

Image from LUMEN - Loyola University Medical Education Network, used with permission.

All rights reserved.
Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2007 and later

Source: Burnsurgery.org
Image used with permission. All rights reserved.
Cutaneous Melanoma Multiple Primary Rules  
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)  
Rules Apply to Cases Diagnosed 1/1/2007 and later

**Unknown if Single or Multiple Melanomas**

*Note:* Melanoma(s) not described as metastasis

**Rule M1**  When it is not possible to determine if there is a **single** melanoma or **multiple** melanomas, opt for a single melanoma 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 Melanoma.

---

**Single Melanoma**

*Note 1:* Melanoma not described as metastasis  
*Note 2:* Includes combinations of in situ and invasive

**Rule M2**  A **single melanoma** is always a single primary. *  
* Prepare one abstract. Use the histology coding rules to assign the appropriate histology code.  
This is the end of instructions for Single Melanoma.

---

**Multiple Melanomas**

Multiple melanomas may be a single primary or multiple primaries  
*Note 1:* Melanoma not described as metastases  
*Note 2:* Includes combinations of in situ and invasive

**Rule M3**  Melanomas in sites with ICD-O-3 **topography** codes that are **different** at the second (Cxxx), third (Cxxxx) or fourth (C44x) character are multiple primaries. **

Jump to  **Equivalent Terms and Definitions**  
Jump to  **Histology Rules**  
**Cutaneous Melanoma Rules**  
For cases diagnosed 2007 and later
Rule M4 Melanomas with different laterality are multiple primaries. **

Note: A midline melanoma is a different laterality than right or left.

Example 1: Melanoma of the right side of the chest and melanoma at midline of the chest are different laterality, 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 Melanomas with ICD-O-3 histology codes that are different at the first (Xxxx), second (xXxx) or third number (xxXx) are multiple primaries. **

Rule M6 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 M7 Melanomas diagnosed more than 60 days apart are multiple primaries. **

Rule M8 Melanomas that do not meet any of the above criteria are abstracted as a single primary. *

Note 1: Use the data item “Multiplicity Counter” to record the number of melanomas abstracted as a single primary.

Note 2: When an invasive melanoma follows an in situ melanoma within 60 days, abstract as a single primary.

Note 3: All cases covered by this rule are the same site and histology.

Note 4: The below examples are not exhaustive.

Example 1: Solitary melanoma on the left back and another solitary melanoma on the left chest.

Example 2: Solitary melanoma on the right thigh and another solitary melanoma on the right ankle.

* 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 Melanomas.
Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)
Rules apply to cases diagnosed 1/1/2007 and later

Single Melanoma or Multiple Melanomas Abstracted as a Single Primary

Rule H1  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 melanoma in the medical record
- PET scan

Note 2: Code the specific histology when documented.

Rule H2  Code the histology from the metastatic site when there is no pathology/cytology specimen from the primary site.

Note: Code the behavior /3.

Rule H3  Code the histology when only one histologic type is identified.

Rule H4  Code the invasive histologic type when there are invasive and in situ components.

Rule H5  Code the histologic type when the diagnosis is regressing melanoma and a histologic type.

Example: Nodular melanoma with features of regression. Code 8721 (Nodular melanoma).

Rule H6  Code 8723 (Malignant melanoma, regressing) when the diagnosis is regressing melanoma.

Example: Malignant melanoma with features of regression. Code 8723.

Rule H7  Code the histologic type when the diagnosis is lentigo maligna melanoma and a histologic type.

Rule H8  Code 8742 (Lentigo maligna melanoma) when the diagnosis is lentigo maligna melanoma.
Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720-8780 (Excludes melanoma of any other site)
Rules apply to cases diagnosed 1/1/2007 and later

Rule H9   Code the most specific histologic term when the diagnosis is melanoma, NOS (8720) with a single specific type.
          Note 1: The specific type for in situ lesions may be identified as pattern, architecture, type, subtype, predominantly, with features of, major, or with ____differentiation
          Note 2: The specific type for invasive lesions may be identified as type, subtype, predominantly, with features of, major, or with ____differentiation.

Rule H10   Code the histology with the numerically higher ICD-O-3 code.

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.