

Solid Tumor Rules

Effective with Cases Diagnosed 1/1/2018 and Forward

Updated November 2020



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Suggested citation: Dickie, L., Johnson, CH., Adams, S., Negoita, S. (November 2020). Solid Tumor Rules. National Cancer Institute, Rockville, MD 20850.

Solid Tumor Rules 2018
Preface
(Excludes lymphoma and leukemia M9590 – M9992)

In Appreciation

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

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

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The 2018 Solid Tumor Rules

Lois Dickie, CTR, Carol Johnson, BS, CTR (Retired), Suzanne Adams, BS, CTR, Serban Negoita, MD, PhD

Preface

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.

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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 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

One site group has been updated for 2021. For cases diagnosed January 1, 2021 and later, the 2018 General Instructions apply to the following revised site listed below:

- Cutaneous Melanoma (Published November 2020)

The 2007 Multiple Primary & Histology rules and the 2007 General Instructions are to be used for cases diagnosed 1/1/2007 to 12/31/2021 for the following site group:

- 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 to 2022 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

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

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How to Navigate the Solid Tumor Rule Modules

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

The following functions will help you maneuver within site groups.

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

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How to Use the Solid Tumor Rules

Note: The rules do not apply to hematopoietic primaries (lymphoma and leukemia) of any site. Use the Hematopoietic & Lymphoid Neoplasm Coding Manual and Database for histologies M9590-M9992.

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

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

STOP

How to Use the Equivalent Terms and Definitions

The Equivalent Terms and Definitions contain the following:

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

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Multiple Primary Rules Do NOT Apply to Tumors Described as Metastases

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

How to Use the Multiple Primary Rules

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

Timing Rules

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

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- 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”.
- No evidence of disease (NED) means complete response to treatment.

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Histologic Type ICD-O-3

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

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

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

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

The North American Association of Central Registries (NAACCR) have released the 2018 Guidelines for ICD-O-3 Histology Code and Behavior Update effective for cases diagnosed 1/1/2018 forward and 2021 Guidelines for ICD-O-3 Histology Code and Behavior Update effective for cases diagnosed 1/1/2021 forward. The updates include:

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

The Solid Tumor Editors recommend coding histology using:

- The 2018 Solid Tumor Rules
- The 2021 Cutaneous Melanoma Solid Tumor Rules
- Updated ICD-O histology codes and terms which can be found at: <https://seer.cancer.gov/icd-o-3/>
- The ICD-O-3.2

When a histology code cannot be identified using the above recommendations, submit a question to [Ask a SEER Registrar](#).

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

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

Ambiguous Terminology

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

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

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Priority Order for Using Documentation to Code Histology

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

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

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

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

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Definitions

Note: Use these terms and definitions for all reportable tumors except lymphoma and leukemia primaries (M9590-9992).

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

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

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

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

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

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

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

Foci: Plural of focus.

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

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

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Multiple primaries: More than one reportable case.

NED: Acronym for “no evidence of disease”; disease free

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

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

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

Recurrence: This term has two meanings:

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

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

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

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

Synchronous: See “Simultaneous”.

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

- Data element: followed by its answer (response).

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

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

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

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

Introduction

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

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

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

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

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

Changes from 2007 MPH Rules

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

1. **NST (No Special Type), mammary carcinoma NST, and carcinoma NST** are the new terms for duct or ductal carcinoma. Previously, it was thought that carcinoma originated in the ducts or lobules of the breast, hence the names duct carcinoma and lobular carcinoma. Current thinking is that carcinoma originates in the “terminal duct lobular unit” therefore the preferred term is NST or carcinoma NST.
2. **Mammary carcinoma** is a synonym for carcinoma no special type (NST)/duct carcinoma not otherwise specified (NOS) **8500**. It will **no longer** be coded as carcinoma NOS **8010**.

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3. **DCIS/Carcinoma NST in situ** has a major classification change.
 - A. Subtypes/variant, architecture, pattern, and features **ARE NOT CODED**. The majority of in situ tumors will be coded to **DCIS 8500/2**.
 - B. It is very important to code the grade of all **DCIS**.
 - i. Code grade as designated in current AJCC Manual, SEER Coding Manual, and COC Coding Manual.
 - ii. The current breast **WHO** edition emphasizes coding the **grade** of tumor rather than the **subtype/variant**.
 - iii. The WHO editions are used internationally by pathologists to keep their nomenclature and histology identification current.
 - iv. Over time, **subtypes/variants** will be diagnosed **less frequently**.
4. The invasive subtype/variant is coded **ONLY** when it comprises **greater than 90%** of the tumor. This change has been implemented in both the WHO and in the CAP protocols.
5. **New codes/terms** are identified by asterisks (*) in the histology table in the Terms and Definitions.
6. Excerpt from the CAP Invasive Breast Protocol (page 17): “A modified list is presented in the protocol based on the most frequent types of invasive carcinomas and terminology that is in widespread usage. The modified list is intended to capture the majority of tumors and reduce the classification of tumors being reported as ‘other.’ The WHO classification is presented for completeness”.

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with; (duct **and** lobular is equivalent to duct **with** lobular)
Note: “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Behavior code /2; DCIS; intraductal; noninfiltrating; noninvasive; carcinoma in situ
- Carcinoma; adenocarcinoma
- De novo; new tumor; frank (obsolete term)
- Duct; ductal; NST (no special type); carcinoma NST; mammary carcinoma
- Mammary; breast
- Majority; major; predominantly; greater than 50%
- Simultaneous; synchronous; existing at the same time; concurrent; prior to first course treatment

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- 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

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

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

Terms and Descriptive Language	Site Term and Code
Inferior inner Inferior medial Lower inner quadrant (LIQ) Lower medial	Lower inner quadrant of breast C503
Superior lateral Superior outer Upper lateral Upper outer quadrant (UOQ)	Upper outer quadrant of breast C504
Inferior lateral Inferior outer Lower lateral Lower outer quadrant (LOQ)	Lower outer quadrant of breast C505
Axillary tail of breast Tail of breast NOS Tail of Spence	Axillary tail of breast C506
12:00 o'clock 3:00 o'clock 6:00 o'clock 9:00 o'clock Inferior breast NOS Inner breast NOS Lateral breast NOS Lower breast NOS Medial breast NOS Midline breast NOS Outer breast NOS Overlapping lesion of breast Superior breast NOS Upper breast NOS	Overlapping lesion of breast C508 <i>Note:</i> This is a <u>single</u> tumor which overlaps quadrants/subsite .

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

Terms and Descriptive Language	Site Term and Code
¾ or more of breast involved with tumor Diffuse (tumor size 998) Entire breast Inflammatory without palpable mass Multiple tumors in different subsites (quadrants) within the same breast	Breast NOS C509 <i>Note:</i> Used for: <ul style="list-style-type: none"> • 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

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

Required Histology Terms	Histology Combination Term and Code
<p>DCIS/duct carcinoma/carcinoma NST 8500</p> <p style="text-align: center;">AND</p> <p>Lobular carcinoma 8520</p> <p><i>Note 1:</i> Both histologies, duct and lobular, must have the same behavior code.</p> <p><i>Note 2:</i> 8522 is used when:</p> <ul style="list-style-type: none"> • 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 <p><i>Example:</i> One tumor with invasive duct CA in LOQ RT breast; second tumor with invasive lobular in UOQ RT breast</p> <p><i>Note 3:</i> Do not use 8522 when the diagnosis is carcinoma NST/duct carcinoma with lobular differentiation. See Histology Rules for instructions on coding differentiation.</p>	<p>Invasive carcinoma NST/duct carcinoma and invasive lobular carcinoma 8522/3</p> <p><i>Note 1:</i> CAP uses the term Invasive carcinoma with ductal and lobular features (“mixed type carcinoma”)</p> <p><i>Note 2:</i> Carcinoma NST includes all subtypes/variants of carcinoma NST.</p> <p>DCIS and in situ lobular carcinoma 8522/2</p> <p><i>Note:</i> The lobular carcinoma includes pleomorphic lobular carcinoma in situ 8519/2.</p>
<p>DCIS/duct carcinoma/carcinoma NST OR any ONE subtype/variant of carcinoma NST</p> <p style="text-align: center;">AND</p> <p>Any histology in Table 3 with exception of</p> <ul style="list-style-type: none"> • Lobular carcinoma 8520 and pleomorphic lobular carcinoma in situ 8519/2* • Paget disease 8540 <p><i>Note 1:</i> Both histologies must have the same behavior code.</p> <p><i>Note 2:</i> See Table 3 for carcinoma NST/duct carcinoma subtypes/variants.</p> <p><i>Note 3:</i> Do not use combination code for duct with lobular differentiation. This is a synonym for carcinoma NST.</p>	<p>Invasive carcinoma NST/duct mixed with other types of invasive carcinoma 8523/3</p> <p>DCIS mixed with other in situ carcinoma 8500/2</p> <p><i>Note:</i> Prior to 2018, DCIS and other in situ was coded 8523/2.</p>

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

Required Histology Terms	Histology Combination Term and Code
<p>Lobular carcinoma</p> <p style="text-align: center;">AND</p> <p>Any histology in Table 3 with exception of</p> <ul style="list-style-type: none"> • Duct carcinoma/carcinoma NST/DCIS (and subtypes/variants) 8500 • Paget disease, in situ and invasive <p><i>Note 1:</i> See Table 3 for carcinoma NST/duct carcinoma subtypes/variants. <i>Note 2:</i> This code does not include lobular and Paget disease. See Multiple Primary Rules. Lobular carcinoma and Paget are separate primaries.</p>	<p>Infiltrating lobular mixed with other types of carcinoma 8524/3</p> <p>In situ lobular mixed with other types of in situ carcinoma 8524/2</p>
<p>Paget disease</p> <p style="text-align: center;">AND</p> <p>Underlying DCIS</p> <p><i>Note:</i> Paget disease is classified as malignant /3 in the ICD-O. Paget disease is coded as in situ /2 ONLY when the pathology states the Paget disease is in situ.</p>	<p>Paget disease (invasive or behavior not specified) and DCIS/intraductal carcinoma 8543/3</p> <p>Paget disease (specified as in situ) and DCIS/intraductal carcinoma 8543/2</p>
<p>Paget disease</p> <p style="text-align: center;">AND</p> <p>Underlying infiltrating duct carcinoma/carcinoma NST and all subtypes/variants of infiltrating duct/carcinoma NST (must be a /3)</p> <p><i>Note:</i> See Table 3 for subtypes/variants of carcinoma NST/duct carcinoma.</p>	<p>Paget disease and infiltrating duct carcinoma 8541/3</p>
<p>Any two invasive carcinoma NST subtypes/variants (percentage not stated) abstracted as a single primary</p> <p><i>Note 1:</i> The diagnosis may be two subtypes/variants and the pathologist may mention the presence of duct/carcinoma NST. Ignore the mention of carcinoma NST. <i>Note 2:</i> See Table 3 for subtypes/variants of carcinoma NST/duct carcinoma.</p>	<p>Adenocarcinoma with mixed subtypes 8255/3</p>

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)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Acinic cell carcinoma 8550	Acinar adenocarcinoma Acinar carcinoma	
Adenoid cystic carcinoma (ACC) 8200	ACC Adenocystic basal cell carcinoma Carcinoma adenoides cysticum Cylindromatous carcinoma	
Adenomyoepithelioma with carcinoma 8983	AME Malignant AME	
Apocrine carcinoma 8401 <i>Note:</i> This is a diagnosis that is EXACTLY apocrine carcinoma , not a carcinoma NST with apocrine features, differentiation, or type .		
Carcinoma NST 8500 <i>Note:</i> Cribriform carcinoma may consist of up to 50% tubular formations. The term cribriform/tubular carcinoma is coded as cribriform carcinoma.	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 Cribriform carcinoma 8201/3 Pleomorphic carcinoma 8022/3

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

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
	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)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
	Invasive carcinoma, NST 8500/3 Invasive carcinoma, type cannot be determined 8500/3 Invasive mammary carcinoma 8500/3 Invasive mammary carcinoma associated with encysted papillary carcinoma 8500/3 Invasive mammary carcinoma NST with lobular features 8500/3 Invasive mammary carcinoma NST with medullary features 8500/3 Invasive mammary carcinoma NST with mucinous features 8500/3 Invasive mammary carcinoma NST with tubulo-lobular variant 8500/3 Invasive mammary carcinoma with apocrine features 8500/3 Invasive mammary carcinoma with cribriform features 8500/3 Invasive mammary carcinoma with tubular features 8500/3 Mammary carcinoma in situ 8500/2 Mammary carcinoma/cancer Non-invasive mammary carcinoma 8500/2	
Glycogen-rich clear cell carcinoma 8315	Glycogen-rich carcinoma	Clear cell carcinoma 8310
Inflammatory carcinoma 8530		
Lipid-rich carcinoma 8314	Lipid-secreting carcinoma	

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

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Lobular carcinoma 8520	Alveolar lobular carcinoma Classic lobular carcinoma Intraductal papilloma with lobular carcinoma in situ 8520/2 Invasive lobular carcinoma, alveolar type/variant 8520/3 Invasive lobular carcinoma, solid type 8520/3 Lobular carcinoma in situ 8520/2 Lobular carcinoma with cribriform features Mixed lobular carcinoma (lobular carcinoma NOS and one or more variants of lobular carcinoma) Invasive pleomorphic lobular carcinoma 8520/3 Solid lobular carcinoma Tubulolobular carcinoma	Pleomorphic lobular carcinoma in situ 8519/2* <i>Note:</i> 8519/2 is a new code for in situ /2 tumors only.
Medullary carcinoma 8510	MC	Atypical medullary carcinoma (AMC) 8513

Table continues on next page

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

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
<p>Metaplastic carcinoma NOS or of no special type (NST) 8575</p> <p><i>Note:</i> 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.</p>	<p>Invasive mammary carcinoma with matrix production</p> <p>Metaplastic carcinoma, mixed epithelial and mesenchymal type</p> <p>Metaplastic carcinoma with mesenchymal differentiation</p> <p>Metaplastic carcinoma with squamous features</p> <p>Metaplastic carcinoma with other types of mesenchymal differentiation</p> <p>Mixed metaplastic carcinoma</p>	<p>Carcinosarcoma 8980/3</p> <p>Fibromatosis-like metaplastic carcinoma 8572</p> <p>Low grade adenosquamous carcinoma 8570</p> <p>Metaplastic carcinoma spindle-cell type/spindle cell carcinoma 8032</p> <p>Metaplastic carcinoma with chondroid differentiation/with osseous differentiation 8571</p> <p>Myoepithelial carcinoma 8982</p> <p>Sarcomatoid carcinoma 8033</p> <p>Squamous cell carcinoma 8070</p>
<p>Mucinous carcinoma 8480</p> <p><i>Note 1:</i> This is a diagnosis that is EXACTLY “mucinous carcinoma,” “mucinous duct carcinoma,” “mucinous DCIS” OR “greater than <u>90%</u> mucinous.” See Histology Rules.</p> <p><i>Note 2:</i> 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.</p>	<p>Colloid carcinoma</p> <p>Mucinous adenocarcinoma</p> <p>Mucoid carcinoma</p>	
Mucoepidermoid carcinoma 8430		
Oncocytic carcinoma 8290		

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

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Paget disease of the nipple with no underlying tumor 8540/3		
Papillary carcinoma 8503	Intraductal papillary carcinoma 8503/2* Intraductal papillary carcinoma with DCIS 8503/2* Invasive papillary carcinoma 8503/3 Papillary carcinoma non-invasive 8503/2* Papillary ductal carcinoma in situ 8503/2*	Encapsulated papillary carcinoma 8504 non-infiltrating/intracystic 8504/2 with invasion 8504/3 Micropapillary carcinoma 8507* Solid papillary carcinoma in situ 8509/2* with invasion 8509/3*
Periductal stromal tumor, low grade 9020/3	Phyllodes tumor, malignant	
Polymorphous carcinoma 8525		
Sarcoma NOS 8800/3 <i>Note:</i> Rhabdomyosarcoma 8900/3 is also a NOS with the following subtypes/variants: Alveolar type rhabdomyosarcoma 8920/3 Embryonal type rhabdomyosarcoma 8910/3 Pleomorphic rhabdomyosarcoma 8901/3		Angiosarcoma 9120/3 Hemangiosarcoma Lymphangiosarcoma 9170/3 Malignant hemangioendothelioma Liposarcoma 8850/3 Leiomyosarcoma 8890/3 Osteosarcoma 9180/3 Rhabdomyosarcoma 8900/3 Alveolar type 8920/3 Embryonal type 8910/3 Pleomorphic 8901/3

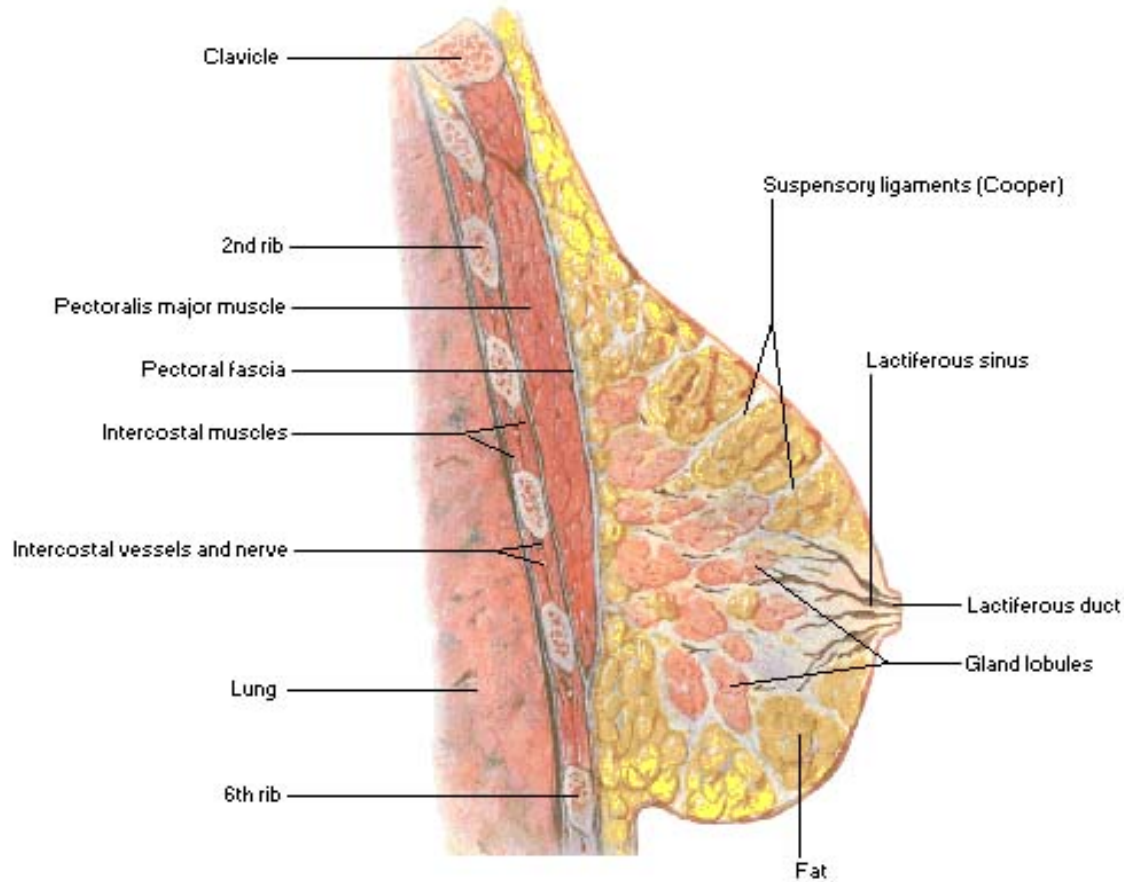
Breast Equivalent Terms and Definitions
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 (Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Specific and NOS/NST Terms and Code	Synonyms	Subtypes/Variants
Sebaceous carcinoma 8410		
Secretory carcinoma 8502	Juvenile breast carcinoma	
Signet ring carcinoma 8490		
Small cell carcinoma 8041	Carcinoid tumor of breast Endocrine carcinoma Neuroendocrine carcinoma, poorly differentiated	Carcinoma with neuroendocrine differentiation/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)

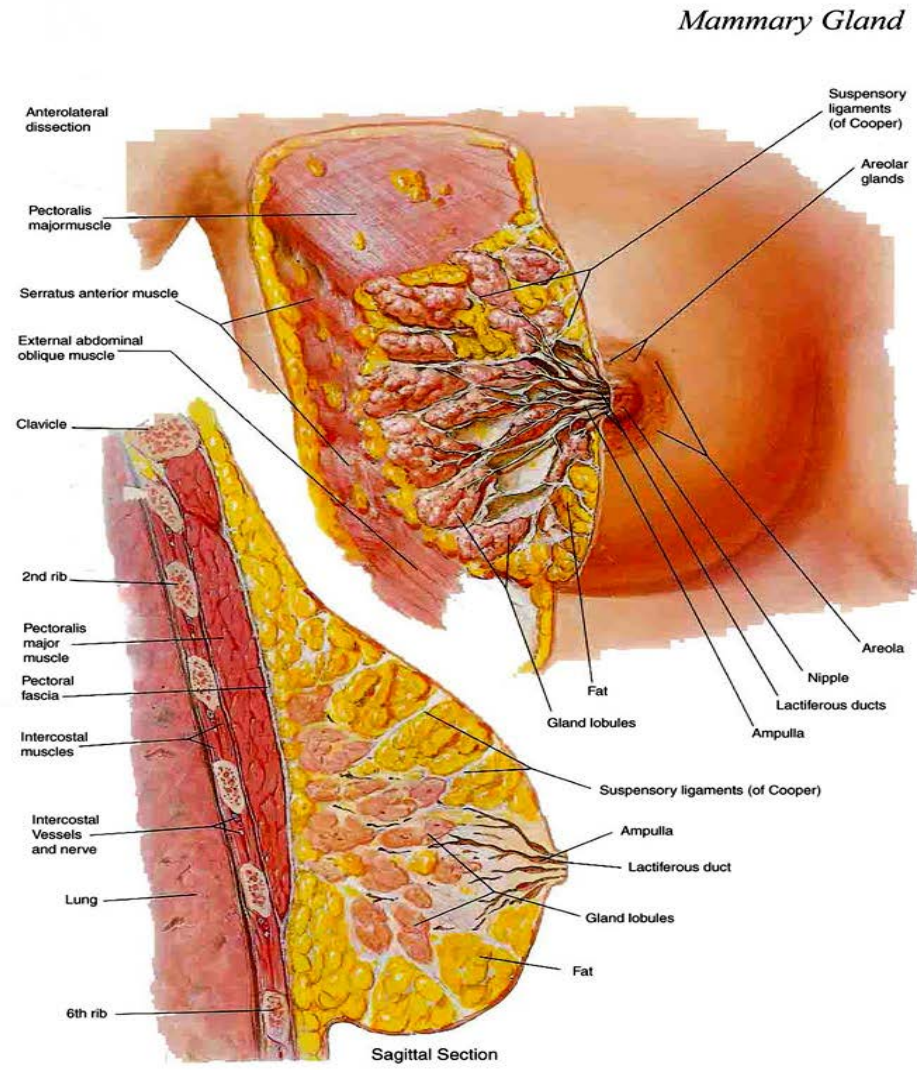
Illustrations



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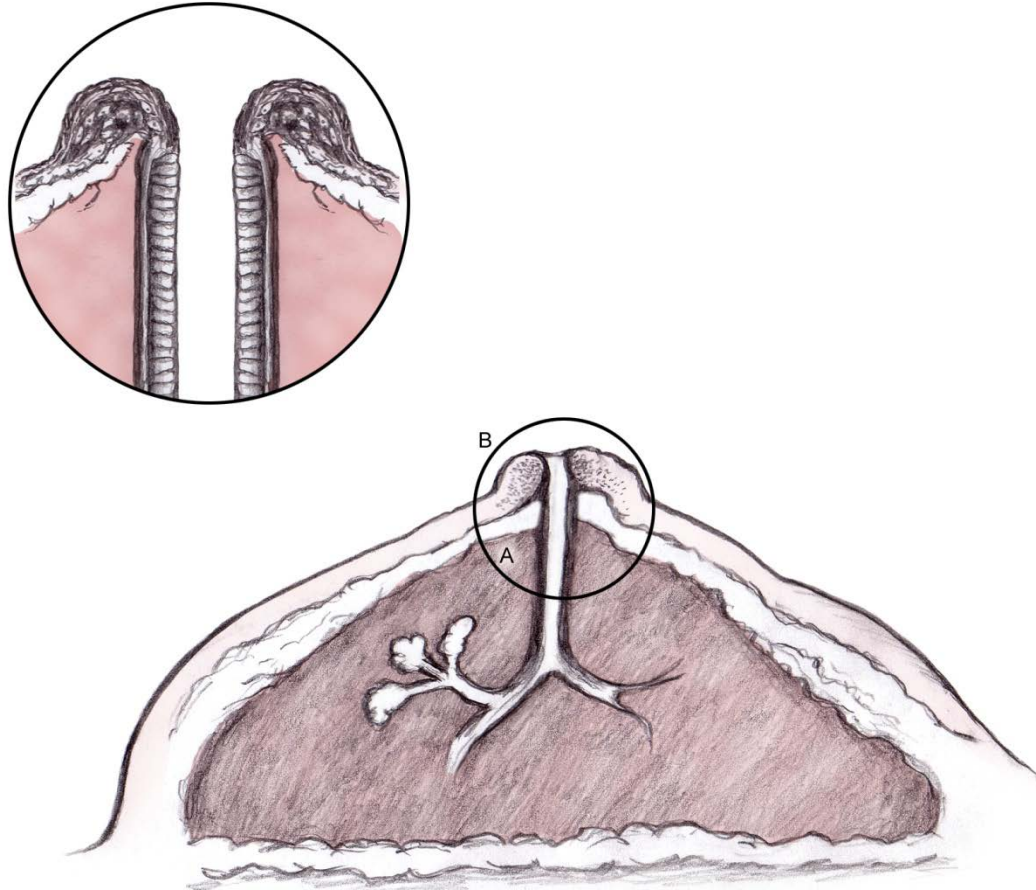
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Breast Equivalent Terms and Definitions
C500-C506, C508-C509
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



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

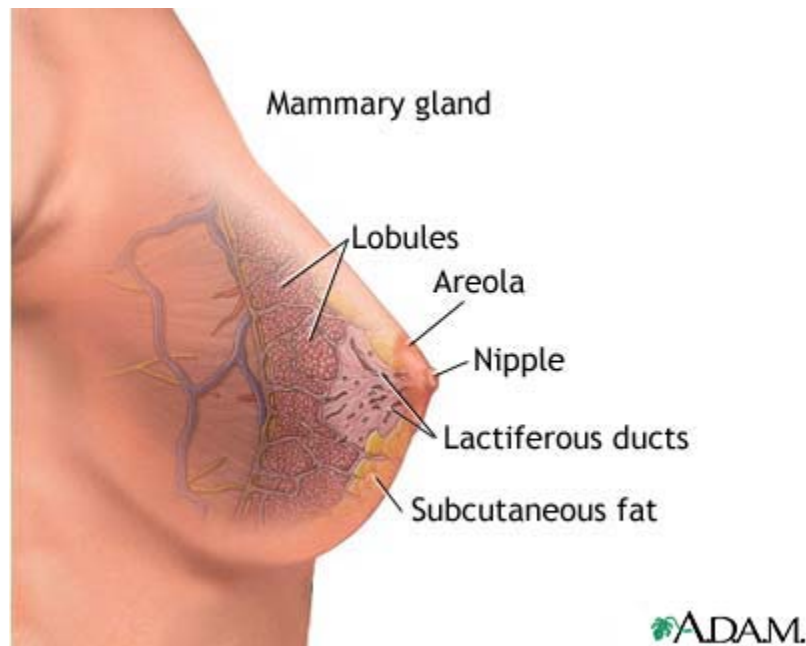


Paget Disease of the nipple. Shows growth pattern of Paget on the pigmented portion of nipple and inside the milk duct opening

Source:

“Image reprinted with permission from eMedicine.com, 2010. Available at: <http://emedicine.medscape.com/article/1101235-overview>”

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



**Breast Equivalent Terms and Definitions
C500-C506, C508-C509**

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

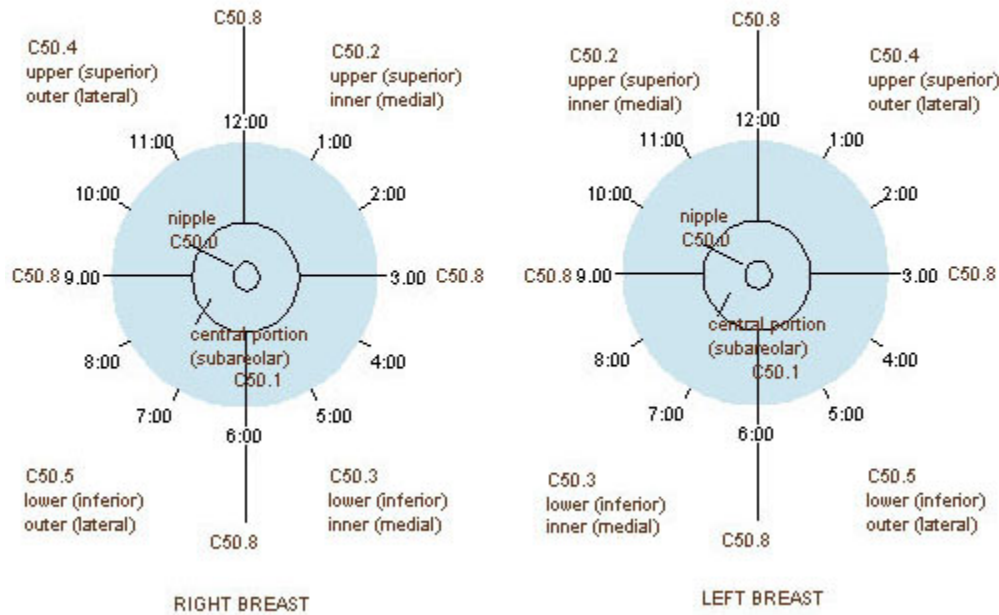
The position of the tumor in the breast may be described as the positions on a clock

The two circles in the graphic are

Innermost circle: Retroareolar (under/behind areola)

Outer circle: Central portion of breast

"Clock" Positions, Quadrants and ICD-O Codes of the Breast



**Breast Multiple Primary Rules
C500-C506, C508-C509**

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

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

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

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

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

Unknown if Single or Multiple Tumors

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

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

Note 2: Examples of cases with minimal information include:

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
 - o Outpatient biopsy with no follow-up information available
 - o 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

ⁱPrepare one abstract. Use the [histology rules](#) to assign the appropriate histology code.

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

Single Tumor

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

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

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

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

Note 1: A single tumor is always a single primary.

Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites/quadrants.

Note 3: The tumor may have in situ and invasive components.

Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor

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

Multiple Tumors

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

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

Note 1: Tumors with site codes that differ at the second or third character are in **different primary sites**; for example, a breast tumor C50x and a colon tumor C18x differ at the second and third character.

Note 2: This rule **does not** include metastases. Metastatic tumors are not used to determine multiple primaries; for example, liver metastases from the breast cancer would not be counted as a second primary.

**Breast Multiple Primary Rules
C500-C506, C508-C509**

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

- Rule M5** Abstract **multiple primaries**ⁱⁱ when the patient has a subsequent tumor after being **clinically disease-free** for **greater than five years** after the original diagnosis or last recurrence.
Note 1: The rules are hierarchical. This rule **only** applies when there is a **subsequent breast tumor**.
Note 2: **Clinically** disease-free means that there was **no evidence** of recurrence on follow-up.
 - Mammograms are NED
 - Scans are NED*Note 3:* When there is a recurrence less than or equal to five years of diagnosis, the “**clock**” starts over. The time interval is calculated from the **date of last recurrence**. In other words, the patient must have been disease-free for greater than five years from the date of the last recurrence.
Note 4: When it is **unknown/not documented** whether the patient had a recurrence, use **date of diagnosis** to compute the time interval.
Note 5: The physician may state this is a **recurrence**, meaning the patient had a previous breast tumor and now has another breast tumor. **Follow the rules**; do not attempt to interpret the physician’s statement.
- Rule M6** Abstract a **single primary**ⁱ when there is **inflammatory carcinoma** in:
 - Multiple quadrants of same breast **OR**
 - Bilateral breasts
- Rule M7** Abstract **multiple primaries**ⁱⁱ when there is **bilateral** breast cancer (both right and left breast).
Note 1: Physician statement of “bilateral breast cancer” should **not be interpreted** as meaning a single primary. The term is not used consistently. The literal definition of bilateral is “cancer in both breasts”.
Note 2: The histologies within each breast may be the same or different.
- Rule M8** Abstract a **single primary**ⁱ when the diagnosis is **Paget disease with synchronous underlying** in situ or invasive carcinoma NST (duct/ductal) or subtypes of duct.
Note: If the underlying tumor is any histology **other than** duct or subtypes of duct, continue through the rules.
- Rule M9** Abstract **multiple primaries**ⁱⁱ when the diagnosis is **Paget disease with underlying** tumor which is NOT duct.
Example: Paget disease of the nipple with underlying lobular carcinoma are multiple primaries.

Breast Multiple Primary Rules
C500-C506, C508-C509

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

Rule M10 Abstract a **single primary**ⁱ when multiple tumors 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**ⁱ when a **ductal** carcinoma occurs **after a combination code** in the same breast. See the following list:

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

Breast Multiple Primary Rules C500-C506, C508-C509

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

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

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

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

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

Note: The same row means the tumors are:

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

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

- On **different rows** in [Table 3](#) in the Equivalent Terms and Definitions
- A combination code in [Table 2](#) and a code from [Table 3](#)

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

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

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

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

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

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

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

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

**Breast Multiple Primary Rules
C500-C506, C508-C509**

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

This is the end of instructions for Multiple Tumors.

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

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

Breast Histology 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
 - o Subtypes/variants of carcinoma NST/duct carcinoma
- Lobular carcinoma NOS
 - o Pleomorphic lobular carcinoma in situ

Ambiguous Terminology

3. Code the specific histology described by **ambiguous terminology** (list follows) **ONLY** when A or B is true:

A. The only diagnosis available is **one histology** term described by ambiguous terminology

- CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
- Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

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

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

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.

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

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.

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

- 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**.¹

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

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

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

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

Rule 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.

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

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 carcinoma²:
- 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).

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

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

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**

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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.

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- 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
- Type; subtype; variant

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**

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

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

Note 1: Rare histologies may not be listed in the table. When a histology term is not found, reference ICD-O and all updates.

Note 2: Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in Table 1, ICD-O or all updates.

Note 3: Behavior codes are listed when the term has only one possible behavior (either a /2 or /3). For histologies which may be either /2 or /3, a behavior code is not listed. Code behavior from pathology.

Column 1 contains specific and NOS histology terms.

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

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

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

Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
<p>Adenocarcinoma 8140</p> <p><i>Note 1:</i> See Histology Rules for instructions on coding adenocarcinoma subtypes/variants arising in a polyp</p> <p><i>Note 2:</i> When the term intestinal adenocarcinoma is used to describe a colon primary, it simply means the appearance is</p>	<p>Adenocarcinoma, NOS</p> <p>Adenocarcinoma/carcinoma in a polyp NOS (now coded to 8140)</p> <p>Adenocarcinoma/carcinoma in adenomatous polyp (now coded to 8140)</p> <p>Adenocarcinoma/carcinoma in polypoid adenoma (now coded to 8140)</p> <p>Adenocarcinoma/carcinoma in serrated adenoma (now coded to 8140)</p> <p>Adenocarcinoma and mucinous carcinoma, mucinous documented as less than 50% of tumor OR percentage of mucinous</p>	<p>Adenoid cystic carcinoma 8200</p> <p>Cribriform comedo-type carcinoma/adenocarcinoma, cribriform comedo-type 8201*</p> <p>Diffuse adenocarcinoma/carcinoma 8145</p> <p>Linitis plastica 8142/3</p> <p>Medullary adenocarcinoma/carcinoma 8510</p> <p>Micropapillary carcinoma 8265*</p> <p>Mucinous/colloid adenocarcinoma/carcinoma 8480</p> <p>Mucoepidermoid carcinoma 8430</p> <p>Serrated adenocarcinoma 8213*</p>

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Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
similar to adenocarcinoma seen in the stomach and is coded to adenocarcinoma NOS 8140	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	Signet ring cell/poorly cohesive adenocarcinoma/carcinoma 8490 Superficial spreading adenocarcinoma 8143 Tubulopapillary carcinoma 8263 Undifferentiated adenocarcinoma/carcinoma 8020
Adenosquamous carcinoma 8560 <i>Note:</i> This code cannot be used for adenocarcinoma subtypes/variants with squamous cell/epidermoid carcinoma	Mixed adenocarcinoma NOS and epidermoid carcinoma Mixed adenocarcinoma NOS and squamous cell carcinoma	

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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Specific and NOS Term and Code	Synonyms for Specific or NOS Term	Subtypes/Variants
Combined small cell carcinoma 8045	Small cell carcinoma mixed with <ul style="list-style-type: none"> • 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	Goblet cell carcinoid 8243
Neuroendocrine carcinoma 8246	NEC	Large cell NEC 8013 Small cell NEC 8041
Neuroendocrine tumor Grade 1 (G1) 8240 <i>Note:</i> When the diagnosis is exactly “carcinoid” it may be a Grade 1 or Grade 2 NET. Default is coding NET Grade 1 8240 .	Carcinoid NOS Low-grade neuroendocrine tumor NET Grade 1 (G1) Well differentiated neuroendocrine tumor	EC cell serotonin-producing NET/enterochromaffin cell carcinoid 8241 Neuroendocrine tumor (NET) Grade 2 (G2) 8249 Somatostatin-producing NET 8156
Sarcoma NOS 8800/3		Angiosarcoma/hemangiosarcoma 9120/3 Leiomyosarcoma 8890/3
Spindle cell carcinoma 8032		
Squamous cell carcinoma 8070	Epidermoid carcinoma NOS Squamous cell carcinoma NOS Squamous cell epithelioma	

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

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

Column 1 lists the **non-reportable** histology term and code for NOS or specific

Column 2 lists the **synonym(s)** for the term

Column 3 lists the **subtype/variant** of the NOS term with the histology code

Column 4 lists the **reason** these histologies are **not reportable**

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

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Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
Familial adenomatous polyposis (FAP) No code	Adenomatous polyposis coli Bussey-Garder polyposis Familial multiple polyposis Familiar polyposis coli Familial polyposis of the colon and rectum Familial polyposis of the gastrointestinal tract Gardner syndrome Multiple adenomatosis		Reportable only when there is cancer in a polyp
Gangliocytic paraganglioma 8683/0			Non-malignant
Gastrointestinal stromal tumor 8936/1	GIST NOS GIST, behavior not specified		Non-malignant
Hyperplastic polyp No code			Non-malignant/no code
Inflammatory or pseudopolyp 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

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Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
L cell glucagon-like peptide and PP/PYY-producing NETs 8152/1*			Non-malignant
Leiomyoma 8890/0			Non-malignant
Lipoma 8850/0			Benign accumulation of fat cells that are circumscribed or encapsulated
Low-grade appendiceal mucinous neoplasm 8480/1 <i>Note:</i> May have low-grade, non-invasive pseudomyxoma peritonei, mucinous implants in peritoneum or beyond	LAMN		Non-malignant
Lynch syndrome No code			Non-malignant/no code
Mesenchymal tumors		Granular cell tumor 9580/0 Hemangioma 9120/0	Non-malignant
Peutz-Jeghers polyp No code	Intraepithelial neoplasia in Peutz-Jeghers polyp(s) Periorificial lentiginosis Peutz-Jeghers polyposis Polyps-and-spots syndrome		Non-malignant/no code

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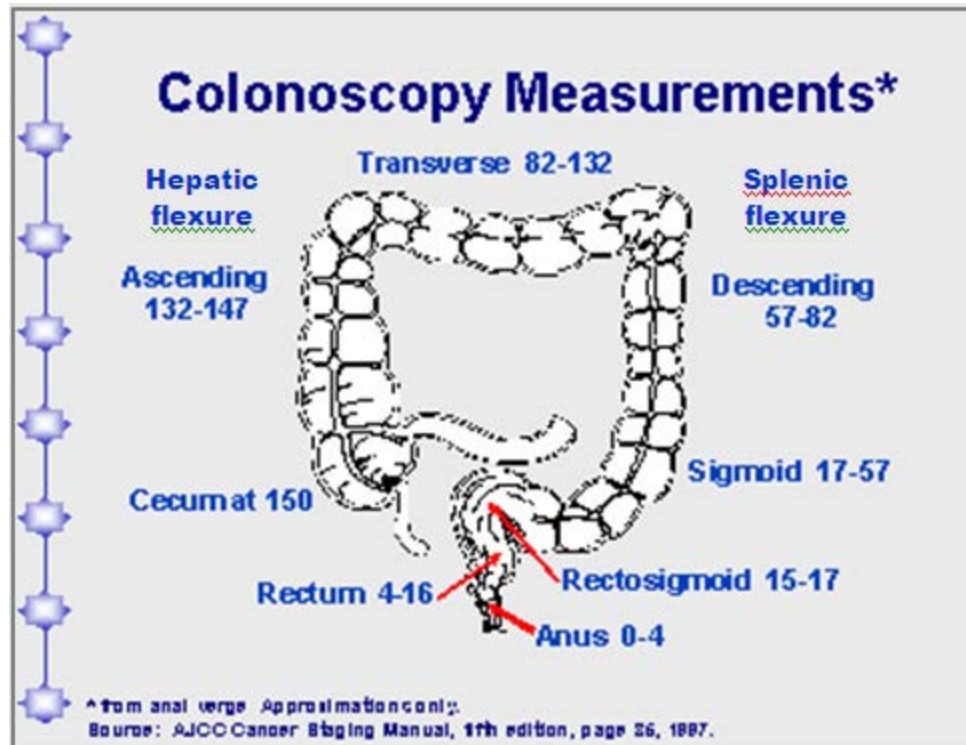
Specific or NOS Term and Code	Synonyms	Subtype/Variant of NOS with Histology Code	Reason not reportable
Pseudomyxoma peritonei (when pathologist does not designate as malignant OR implants are benign) 8480/1			Non-malignant. When both implants and site of origin are benign, the case is not reportable.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

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

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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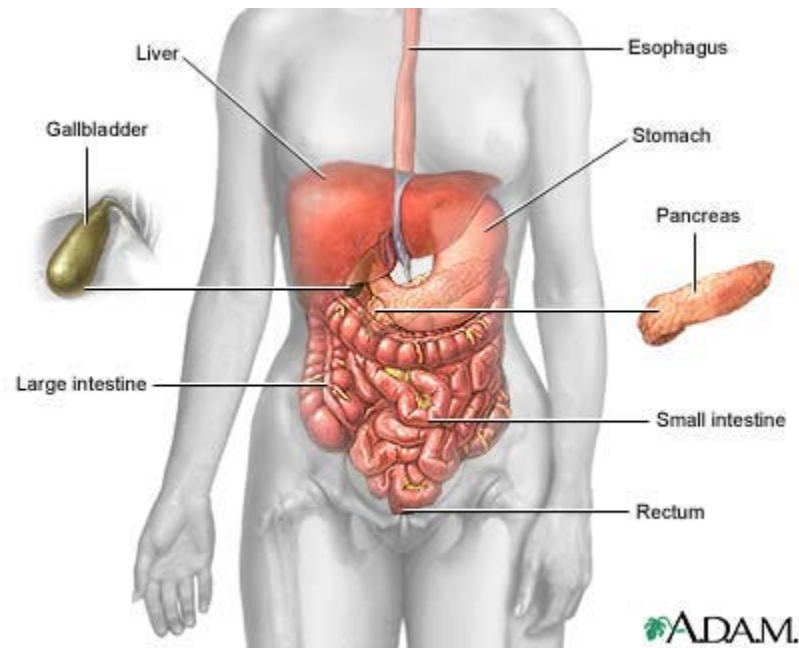
Illustrations

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



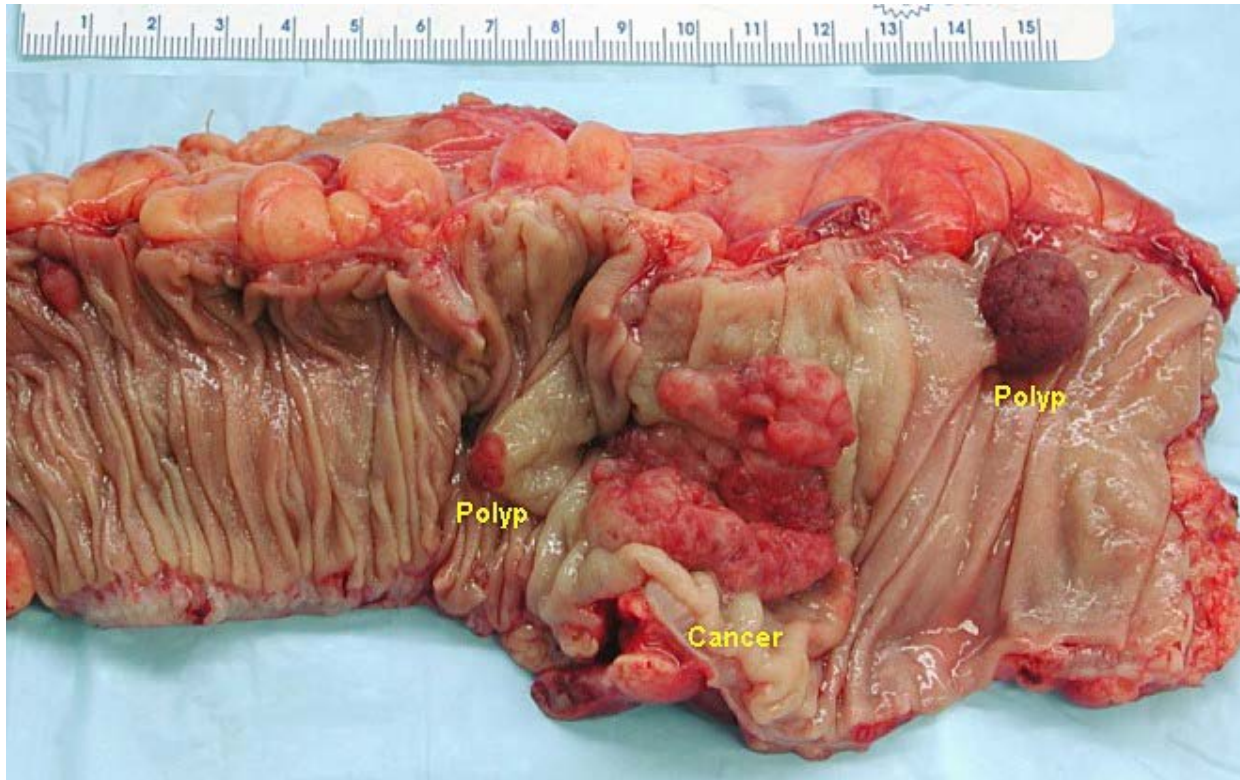
Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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GI System



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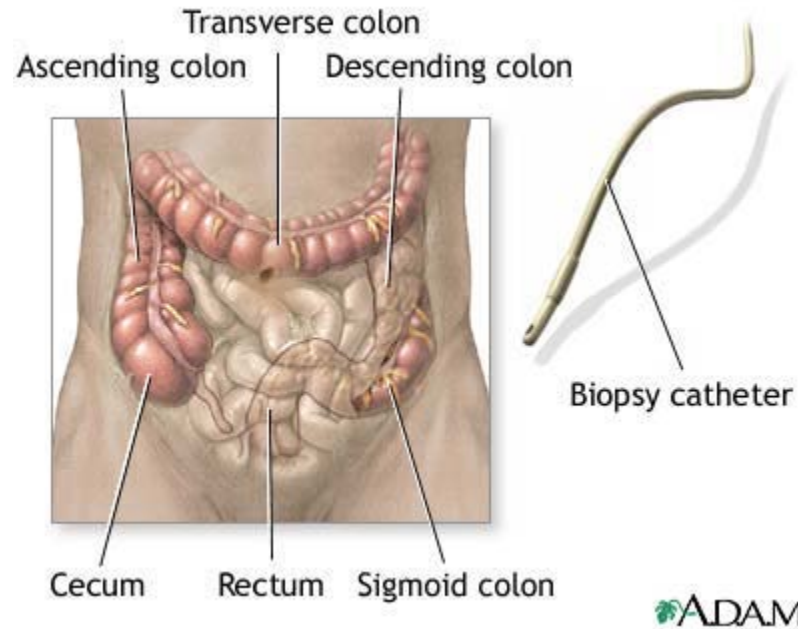
Polyps and de novo or “frank” adenocarcinoma in colon



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

Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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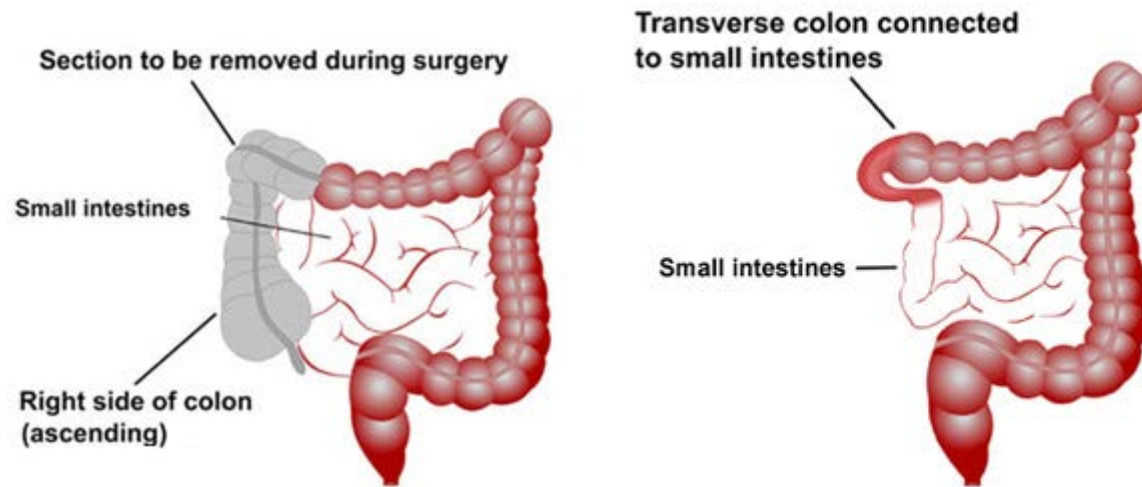
Large intestine; snare instrument to remove polyps



Colon, Rectosigmoid, and Rectum Equivalent Terms and Definitions
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Colon Surgery: Hemicolectomy

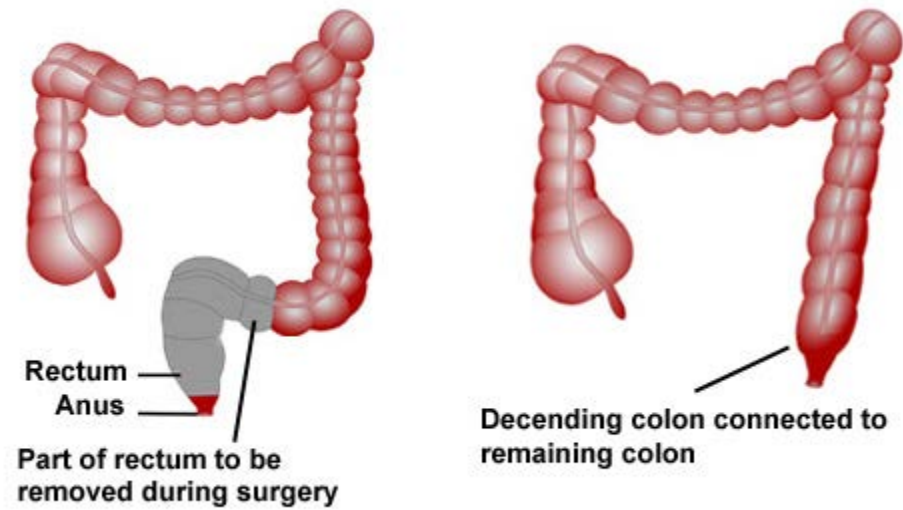
The primary treatment for colon cancer is surgery. Part of the large bowel and the surrounding lymph nodes are removed. The remaining bowel is then joined together (anastomosis).



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

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Rectal Surgery



Colon, Rectosigmoid, and Rectum Multiple Primary Rules
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Note 1: These rules are **NOT** used for tumor(s) described as metastases. Metastatic tumors include but are not limited to:

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

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

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

Unknown if Single or Multiple Tumors

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

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

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

Note 2: Examples of cases with minimal information include

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

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

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

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Single Tumor

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

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

Note 1: A single tumor is always a single primary.

Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.

Note 3: The tumor may have in situ and invasive components.

Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor

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

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
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(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.

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Rule M4 Abstract **multiple primaries**ⁱⁱ when there are separate, non-contiguous tumors in sites with ICD-O site codes that **differ** at the second **CXxx** and/or third **CxXx** 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 **C50x** and colon **C18x**

Example 2: Colon **C18x** and rectum **C209** (This does not include FAP- see earlier rules)

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

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

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

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

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

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

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

- One tumor is a **NOS** and the other is a **subtype/variant** of that NOS **OR**
- The subsequent tumor occurs **greater than 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.

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

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**ⁱ 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**ⁱⁱ when there are separate, non-contiguous tumors in sites with ICD-O site codes that **differ** at the fourth character **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**ⁱⁱ when the patient has a subsequent tumor after being **clinically disease-free** for **greater than one year** after the original diagnosis or last recurrence.

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

- Colonoscopies are NED
- Scans are NED

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

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

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**ⁱ when synchronous, separate/non-contiguous tumors are on **the same row in [Table 1](#)** in the Equivalent Terms and Definitions.

Note: The same row means the tumors are:

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

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

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

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

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

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

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

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

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

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

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

Colon, Rectosigmoid, and Rectum Multiple Primary Rules
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

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

Note 2: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression.

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

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

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

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

This is the end of instructions for Multiple Tumors.

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

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

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

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed prior to neoadjuvant treatment.
Note 1: Histology changes 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
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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)

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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

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

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

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

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

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

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

List of Ambiguous Terminology

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

Colon, Rectosigmoid, and Rectum Histology Rules
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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
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- 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
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(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**
 - Less than or equal to 100 polyps are identified **OR**
 - The exact number of polyps is unknown/not documented

Note 1: **Do not use** this code for a malignancy in a **single polyp** (adenoma) or for a de novo (frank) malignancy.

Note 2: Use this rule **ONLY** for **adenocarcinoma NOS** in multiple polyps.

Rule 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.

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 C411 and maxilla C410, 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
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Equivalent or Equal Terms

These terms can be used interchangeably:

- Adenocarcinoma; adenocarcinoma NOS; carcinoma; carcinoma NOS
- And; with
Note: “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**
- Contiguous; continuous
- 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**
- Type; subtype; variant

Head and Neck Equivalent Terms and Definitions
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Terms that are NOT Equivalent or Equal

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

- **Component** is not equivalent to **subtype/type/variant**
Note: Component is **only** coded when the pathologist specifies the component as a second **carcinoma**
- **P16 positive** is not equivalent to **HPV positive**
- **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
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- 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

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

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

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

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

Head and Neck Equivalent Terms and Definitions
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Table Index

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<u>Table 1</u>	Tumors of Nasal Cavity C300 Paranasal Sinuses C310-C313, C318, C319
<u>Table 2</u>	Tumors of Nasopharynx C110, C111 (posterior wall of nasopharynx only), C112, C113, C118, C119
<u>Table 3</u>	Pyiform Sinus C129 Tumors of Hypopharynx C130-C132, C138, C139 Larynx C320-C323, C328, C329 Trachea C339 and Parapharyngeal Space C139
<u>Table 4</u>	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
<u>Table 5</u>	Tumors of Oropharynx C100-C104, C108 C109 Base of Tongue C019 Tonsils C090, C091, C098, C099 Adenoids/pharyngeal tonsil only C111
<u>Table 6</u>	Tumors of Salivary Glands C079, C080, C081, C088, C089
<u>Table 7</u>	Tumors of Odontogenic and Maxillofacial Bone (Mandible C411 , Maxilla C410)
<u>Table 8</u>	Tumors of Ear C301 and External auditory canal C442
<u>Table 9</u>	Paraganglioma of Carotid body, Larynx, Middle Ear, Vagal nerve C479
<u>Table 10</u>	Paired Sites

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 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)

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

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)

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

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)

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

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

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

Table 2: Tumors of Nasopharynx

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

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

Note 1: The **nasopharynx** is the upper part of the pharynx. It is above the soft palate and extends to the nasal passages.

Note 2: Nasopharyngeal tumors are usually assigned to the subsite in which they occur.

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the [Hematopoietic Database](#).

Note: Hematopoietic tumors are common to the nasopharynx.

Column 1 contains specific and NOS histology terms.

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

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

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

Table begins on next page.

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

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

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

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

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

Note 1: The **hypopharynx** is in the inferior position of the three segments of pharynx. The hypopharynx links the oropharynx to the esophagus, lower part of the pharynx. The pyriform sinus is located in the hypopharynx.

Note 2: The **larynx** is only 1 1/2 inches. It is inferior to the hyoid bone and tongue. It is anterior to the esophagus.

Note 3: The **trachea** starts where larynx ends and continues down the middle of the neck anterior to the esophagus.

Note 4: The **parapharyngeal space** is an equivalent of the lateral pharyngeal space which includes the soft tissue, vessels and skeletal muscles supporting the mechanics of the pharynx. Code the specific site when the soft tissue, vessel, or skeletal muscle is documented. When specific information is not available/not documented, code hypopharynx NOS, C139.

Note 5: These primary sites are mostly composed of muscle and cartilage, but the most common tumors arise from the epithelial lining of the structures (squamous cell carcinoma, for example).

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)

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.

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenoid cystic carcinoma 8200	ACC (rare)	
Chondrosarcoma 9220	Chondrosarcoma grade 2/3 Chondrosarcoma NOS	
Liposarcoma 8850	Atypical lipomatous tumor Well-differentiated liposarcoma	
Squamous cell carcinoma (SCC) 8070	Epidermoid carcinoma Squamous cell carcinoma NOS	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
Well-differentiated neuroendocrine carcinoma 8240	Carcinoid Neuroendocrine carcinoma grade 1	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

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)

C061 Vestibule of mouth, alveolar sulcus, buccal sulcus, labial sulcus

C062 Retromolar area, retromolar triangle, retromolar trigone

C068 Overlapping lesion of other and unspecified parts of mouth

C069 Mouth NOS, buccal cavity, oral cavity, oral mucosa, minor salivary gland NOS

Note: There is no ICD-O site code for minor salivary glands. Many minor salivary glands are located in the lips, inner cheek (buccal mucosa) and there are extensive minor salivary glands in the linings of the mouth and throat. Code to the site in which the salivary gland is located.

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the [Hematopoietic Database](#).

Note: Hematopoietic tumors are common to the oral cavity.

Column 1 contains specific and NOS histology terms.

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

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

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

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Kaposi sarcoma 9140	Kaposi disease	
Mucoepidermoid carcinoma 8430	Mucoepidermoid tumor	
Myofibroblastic sarcoma 8825	Myofibrosarcoma	
Oral mucosal melanoma 8720		
Squamous cell carcinoma 8070	Squamous carcinoma Squamous cell carcinoma NOS	Acantholytic squamous cell carcinoma 8075

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

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

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

Tonsils:

C090 Tonsillar fossa

C091 Tonsillar pillar

C098 Overlapping lesion of tonsil

C099 Tonsil NOS

C111 Adenoids/pharyngeal tonsil (does not include posterior wall of nasopharynx)

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the [Hematopoietic Database](#).

Column 1 contains specific and NOS histology terms.

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

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

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

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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)

Specific or NOS Term and Code	Synonyms	Subtypes/Variants
Adenoid cystic carcinoma 8200		
Polymorphous adenocarcinoma 8525	Cribriform adenocarcinoma Polymorphous low-grade adenocarcinoma Terminal duct carcinoma	
Squamous cell carcinoma 8070		Keratinizing squamous cell carcinoma 8071 Non-keratinizing squamous cell carcinoma 8072 Squamous cell carcinoma HPV-negative 8086* Squamous cell carcinoma HPV-positive 8085* <i>Note:</i> 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.

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

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

Table 6: Tumors of Salivary Glands

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

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

For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the [Hematopoietic Database](#).

Note: Hematopoietic neoplasms are common in the major salivary glands.

Column 1 contains specific and NOS histology terms.

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

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

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

Note 1: **Salivary duct carcinoma** was **assigned** code **8500** because it resembles high-grade duct carcinoma as found in the breast.

These tumors are very aggressive. Code **8500 only** when the diagnosis is **exactly salivary duct carcinoma**.

Note 2: Assign code **8140** when the diagnosis is **salivary gland adenocarcinoma**.

Table begins on next page

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

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

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

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

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

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

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

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)

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

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

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

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

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 8: Tumors of Ear and External Auditory Canal

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.

Specific or NOS Term and Code	Synonyms
Ceruminous adenocarcinoma 8420 <i>Note:</i> The primary site is ceruminous gland C442.	Ceruminal adenocarcinoma
Endolymphatic sac tumor 8140 <i>Note:</i> The endolymphatic sac is located within the inner ear C301.	Adenocarcinoma Heftner tumor Low-grade papillary adenocarcinoma of endolymphatic sac origin
Squamous cell carcinoma of the middle ear 8070 <i>Note:</i> This neoplasm arises in the squamous epithelium within the middle ear C301.	SCC Squamous carcinoma Squamous cell carcinoma NOS

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 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

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

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

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

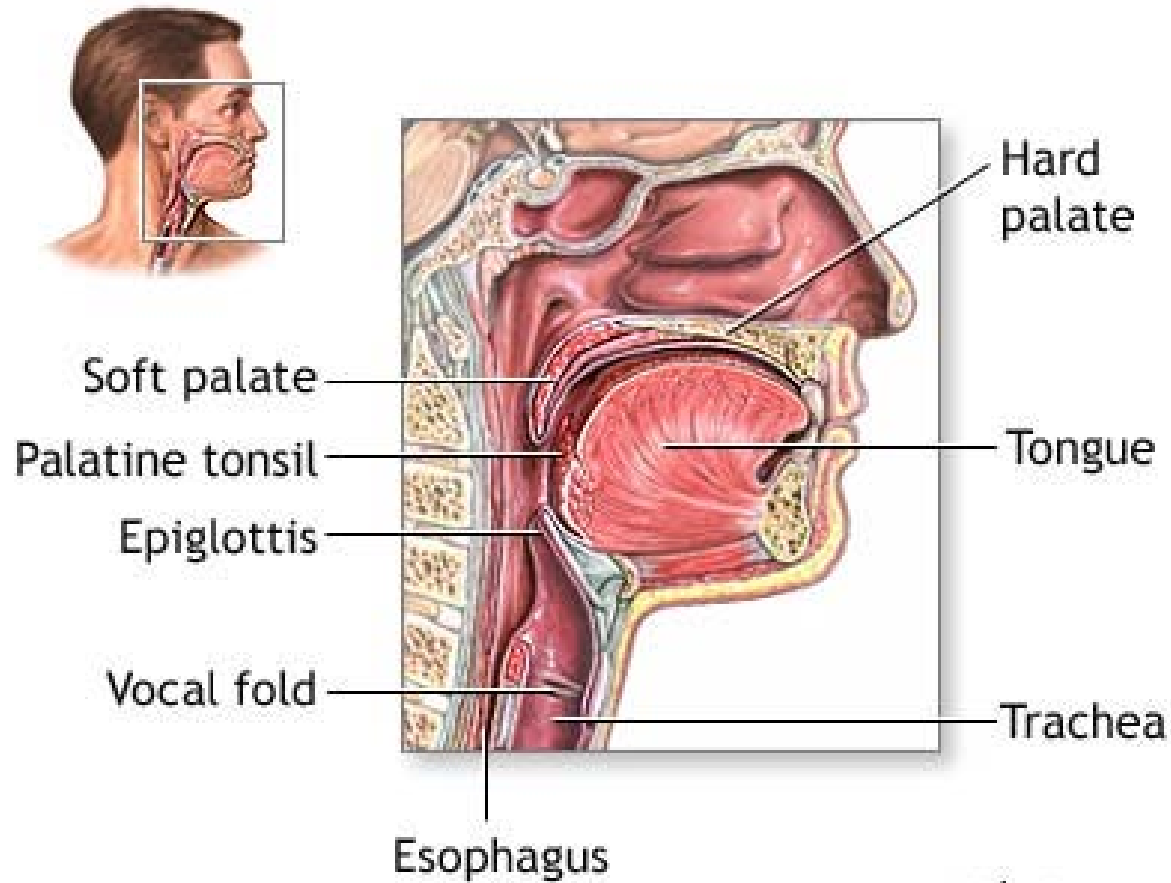
Table 10: Paired Sites

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

Paired Sites	Site Code
Frontal sinus	C312
Maxillary sinus	C310
Middle ear	C301
Nasal cavity (excluding nasal cartilage, nasal septum)	C300
Tonsil	C09_
Parotid gland	C079
Skin of External Ear	C442
Sublingual gland	C081
Submandibular gland	C080

Head and Neck Equivalent Terms and Definitions
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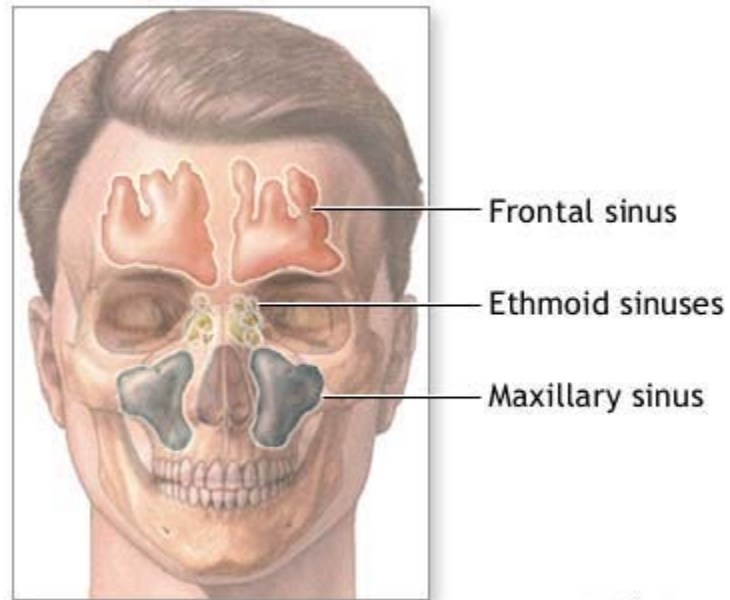
Illustrations



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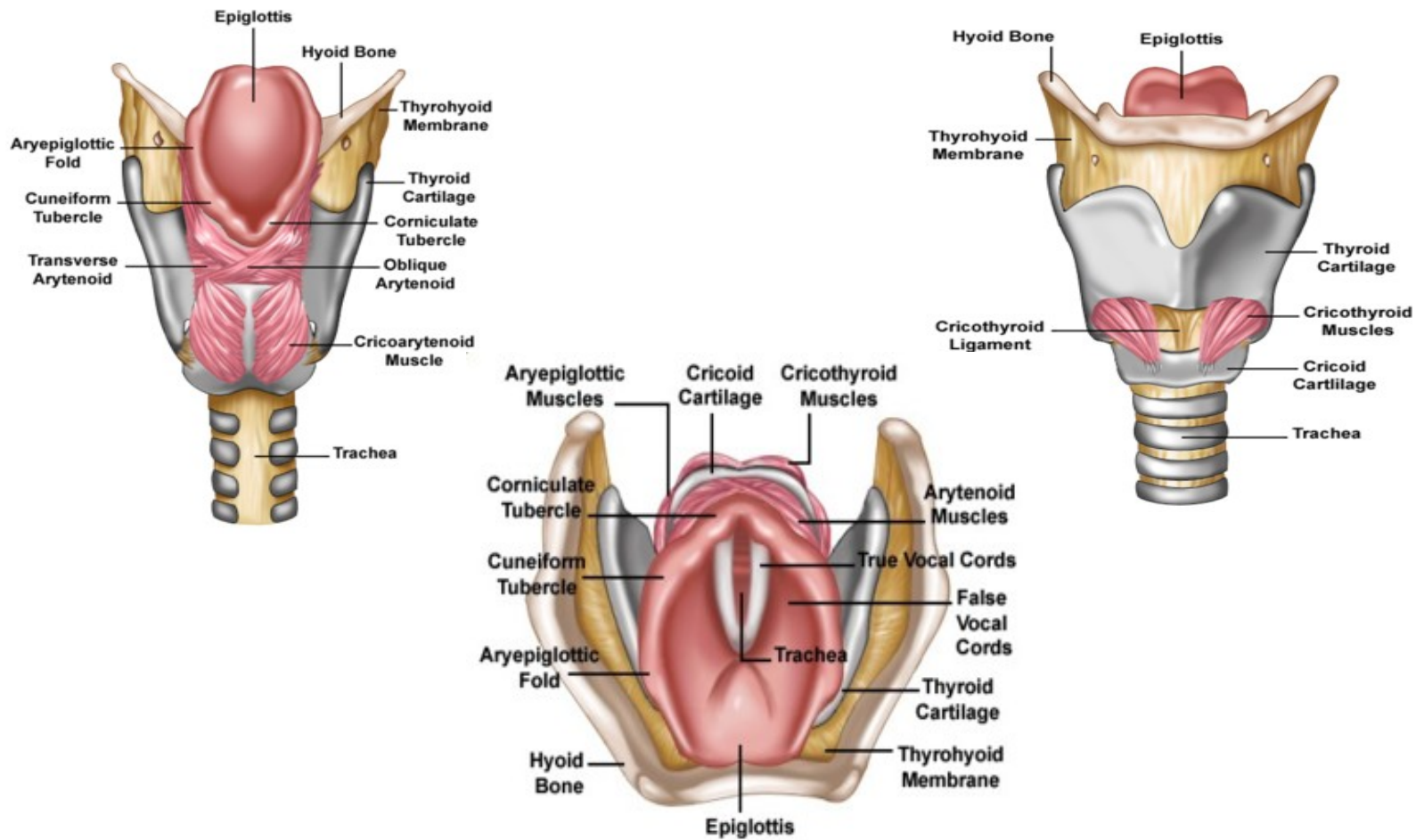
Head and Neck Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



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Nasal Sinuses

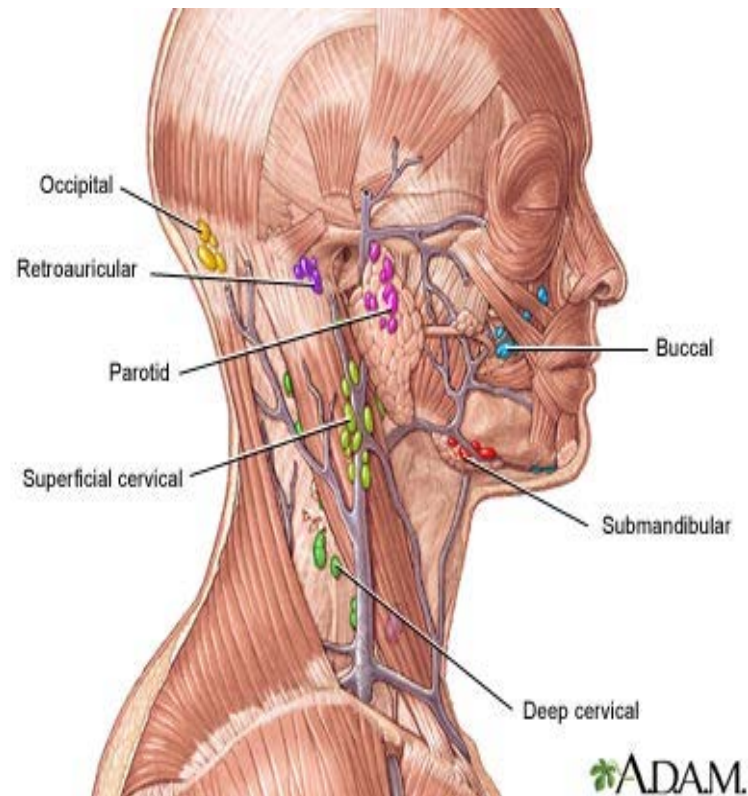
Head and Neck Equivalent Terms and Definitions
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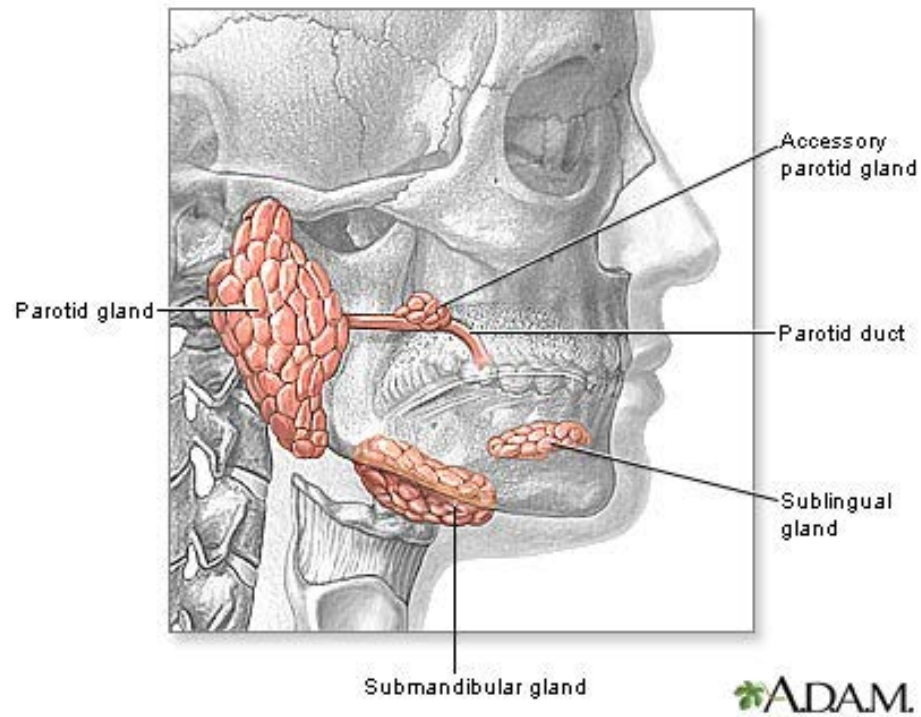
Larynx

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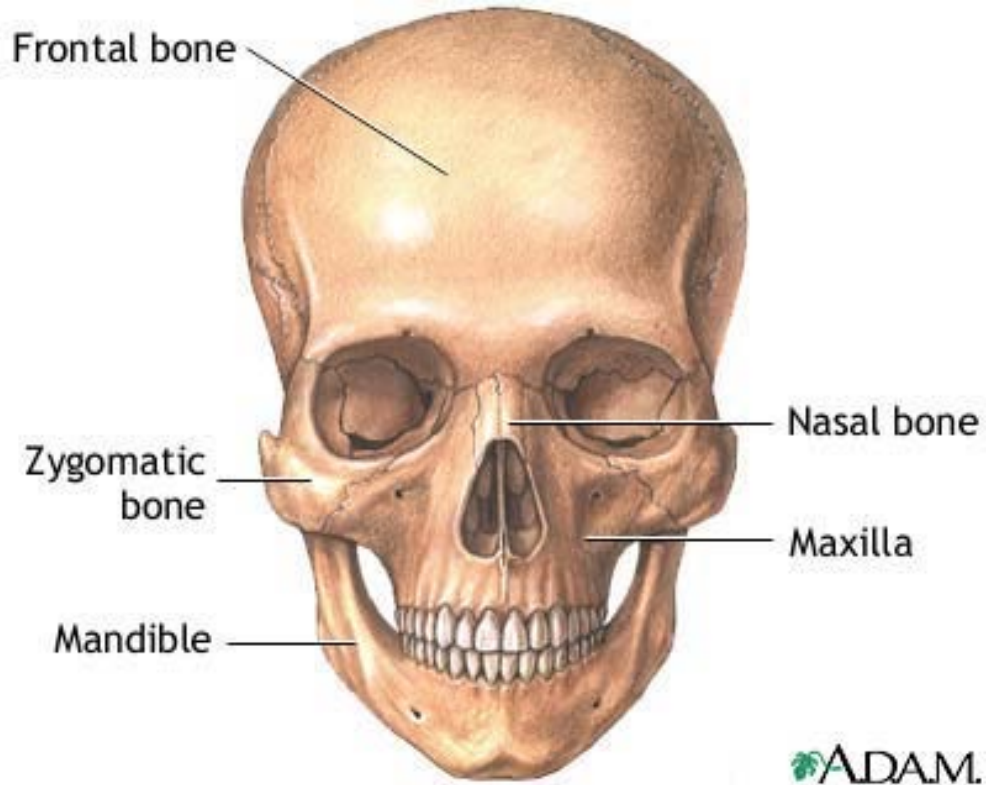
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Head and Neck Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

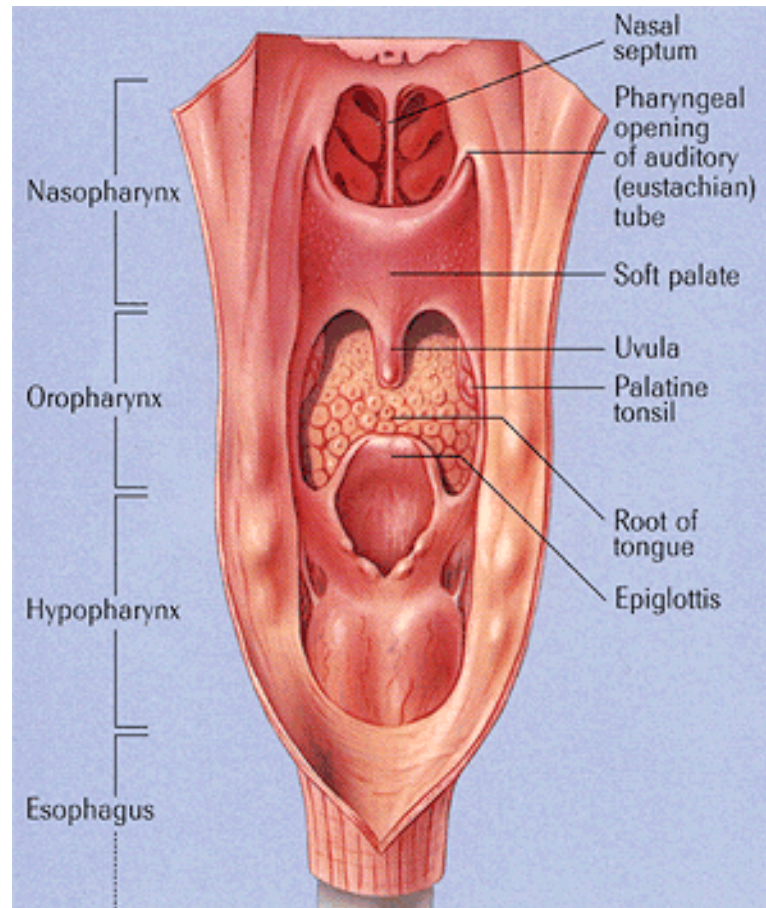


Image made available by a generous grant from Bristol-Myers Squibb

Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, 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**ⁱ when it is not possible to determine if there is a **single tumor or multiple tumors**.

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

Note 2: Examples of cases with minimal information include

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

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

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

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

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

Head and Neck Multiple Primary Rules
C000-C148, C300-C339, C410, C411, 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**ⁱⁱ when there are separate/non-contiguous tumors in any two of the following sites:

- Glottis **C320 AND/OR** supraglottis **C321 AND/OR** subglottis **C322 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 CxxX. Use this rule **ONLY** for the primary sites listed.

Rule M4 Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are present in sites with ICD-O **site** codes that **differ** at the second CXxx, and/or third characters CxXx.

Note 1: Use this rule only for **multiple tumors**.

Note 2: Timing is irrelevant.

Note 3: Histology is irrelevant.

Rule M5 Abstract **multiple primaries**ⁱⁱ when there are separate/non-contiguous tumors on both the **right** side and the **left** side of a paired site.

Note 1: See [Table 10](#) for a list of **paired sites**.

Note 2: Use this rule only for **multiple tumors**.

Note 3: Timing is irrelevant.

Note 4: Histology is irrelevant.

Rule M6 Abstract **multiple primaries**ⁱⁱ when the patient has a subsequent tumor after being **clinically disease-free** for **greater than five years** after the original diagnosis or last recurrence.

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

- Scopes are NED
- Scans are NED

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

Note 3: When it is **unknown/not documented** whether the patient had a recurrence, use **date of diagnosis** to compute the time interval.

Note 4: The physician may state this is a **recurrence**, meaning the patient had a previous head and neck tumor and now has another head and neck tumor. **Follow the rules**; do not attempt to interpret the physician’s statement.

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

- Rule M7** Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3 of the appropriate site table ([Tables 1-9](#)) in the Equivalent Terms and Definitions. Timing is irrelevant.
Note: The tumors may be subtypes/variants of the **same** or **different** NOS histologies.
- **Same NOS:** Alveolar rhabdomyosarcoma 8920/3 and embryonal rhabdomyosarcoma 8910/3 are both subtypes of rhabdomyosarcoma 8900/3 but are distinctly different histologies. Abstract multiple primaries.
 - **Different NOS:** Colloid-type adenocarcinoma 8144 is a subtype of adenocarcinoma NOS 8140; Spindle cell squamous cell carcinoma 8074 is a subtype of squamous cell carcinoma 8070. They are distinctly different histologies. Abstract multiple primaries.
- Rule M8** Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are on **different rows** in the appropriate site table ([Tables 1-9](#)) in the Equivalent Terms and Definitions. Timing is irrelevant.
Note: Each row in the table is a **distinctly different** histology.
- Rule M9** Abstract a **single primary**ⁱ (the invasive) when an **in situ** tumor is diagnosed **after** an **invasive tumor in the same primary site**.
Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS. See [Tables 1-9](#) in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.
Note 3: 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**ⁱ (the invasive) when an **invasive tumor** is diagnosed **less than or equal to 60 days after** an **in situ tumor in the same primary site**.
Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: The tumors may be an **NOS** and a **subtype/variant** of that NOS.

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**ⁱⁱ when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor.

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

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

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

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

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

Note: The same row means the tumors are:

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

Rule M13 Abstract a **single primary**ⁱ when none of the previous rules apply.

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

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

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

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

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed prior to neoadjuvant treatment.
Note 1: Histology changes 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 biopsy or resection of primary site** (in priority order)

- A. Addendum(s) and/or comment(s)
- B. Final diagnosis / synoptic report as required by CAP
- C. CAP protocol

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

Note 2: The pathologist’s diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.

Note 3: The CAP protocol is a checklist which:

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

2. Cytology of primary site (fine needle aspirate (FNA))

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

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.

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

3. Code the specific histology described by **ambiguous terminology** (list follows) **ONLY** when A or B is true:

A. The only diagnosis available is **one histology** term described by ambiguous terminology

- CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
- Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

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

B. There is a **NOS histology and a more specific** (subtype/variant) described by ambiguous terminology

- Specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.) **OR**
- Patient is receiving treatment based on the specific histology described by ambiguous term

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

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

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

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

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

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

- Architecture
- Foci; focus; focal
- Pattern

Head and Neck Histology Rules
C000-C148, C300-C339, C410, C411, 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/endolymphatic sac tumor **8140** and a subtype/variant of adenocarcinoma
- Ameloblastic carcinoma primary type **9270** and a subtype variant of ameloblastic carcinoma primary type
- Chondrosarcoma grade 2/3 **9220** and a subtype/variant of chondrosarcoma grade 2/3
- Neuroendocrine carcinoma **8246** and a subtype/variant of neuroendocrine carcinoma
- Odontogenic carcinosarcoma **8980** and a subtype/variant of odontogenic carcinosarcoma
- Sarcoma **8800/3** and a subtype/variant of sarcoma
- Squamous cell carcinoma **8070** and subtype/variant of squamous carcinoma
- Well differentiated neuroendocrine carcinoma **8240** and a subtype/variant of well differentiated neuroendocrine carcinoma

Note: See [Tables 1-9](#) in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

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

Code the histology according to the rule that fits the case

Jump to [Equivalent Terms and Definitions](#)

Jump to [Multiple Primary Rules](#)

Solid Tumor Rules
Updated November 2020

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

Introduction

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

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

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

Note 3: **Renal cell carcinoma (RCC) 8312** is a **group term** for glandular (adeno) carcinoma of the kidney. Approximately 85% of all malignancies of the kidney C649 are RCC or subtypes/variants of RCC.

- See [Table 1](#) for renal cell carcinoma subtypes/variants.
- Clear cell renal cell carcinoma (ccRCC) 8310 is the most common subtype/variant of RCC.

Note 4: **Transitional cell carcinoma** rarely arises in the kidney C649. Transitional cell carcinoma of the upper urinary system usually arises in the renal pelvis C659. Only code a transitional cell carcinoma for kidney in the **rare** instance when **pathology confirms** the tumor originated in the kidney.

Note 5: For those sites/histologies which have recognized **biomarkers**, the biomarkers are most frequently used to target treatment. Biomarkers may identify the histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Changes from 2007 Rules

These changes are effective with cases diagnosed 1/1/2018 and later. WHO Classification of Tumors of the Urinary System and Male Genital Organs was published in 2016.

1. 2007 Rules instruct “Code the histology from the most representative specimen.” For all sites except breast and CNS, 2018 Rules instruct “Code the most specific histology from biopsy or resection. When there is a discrepancy between the biopsy and resection

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(two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).”

2. **New histology terms and codes were included** (identified by asterisks (*) in the histology table in the Terms and Definitions).
 - A. Histologies with terms that indicate they are **hereditary** (hereditary leiomyomatosis and renal cell carcinoma syndrome–associated RCC **8311**)
 - B. Histologies with **genetic anomalies** (succinate dehydrogenase–deficient RCC)
3. Some histologies are rare and are not listed in the tables; refer to ICD-O and all updates.
Note: Renal cell spindle cell carcinoma 8318 is no longer a recommended term.

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
Note: “And” and “with” are used as synonyms when describing multiple histologies within a single tumor.
- Multifocal; multicentric
- Carcinoma; adenocarcinoma
- 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

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Terms that are NOT Equivalent or Equal

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

- **Component** is not equivalent to **subtype/type/variant**
Note: Component is only coded when the pathologist specifies the component as a second **carcinoma**
- **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.

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NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants
Nephroblastoma 8960	Wilms tumor	
Neuroendocrine tumor (NET) 8240	Carcinoid [OBS] Well-differentiated neuroendocrine tumor	Large cell neuroendocrine carcinoma/tumor 8013 Small cell neuroendocrine carcinoma 8041
Renal cell carcinoma NOS 8312 <i>Note 1:</i> WHO, IARC, and CAP agree that sarcomatoid carcinoma is a pattern of differentiation, not a specific subtype, of renal cell carcinoma. <i>Note 2:</i> Sarcomatoid is listed in the CAP Kidney protocol under the header “features.”	RCC Sarcomatoid carcinoma Sarcomatoid renal cell carcinoma Succinate dehydrogenase-deficient renal cell carcinoma (SDHD) Unclassified renal cell carcinoma	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 <i>Note:</i> The 2016 WHO 4 th 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* <i>Note:</i> 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* <i>Note:</i> This is a new term (previously called renal spindle cell carcinoma).

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

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

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Table 2: Neoplasms which are Not Reportable

Column 1 lists the not reportable histology **term** and **code**. Not all of the non-reportable neoplasms have codes.
Column 2 lists **synonyms** for the term in column 1. Synonyms have the same histology code (if applicable) as listed in column 1.

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

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

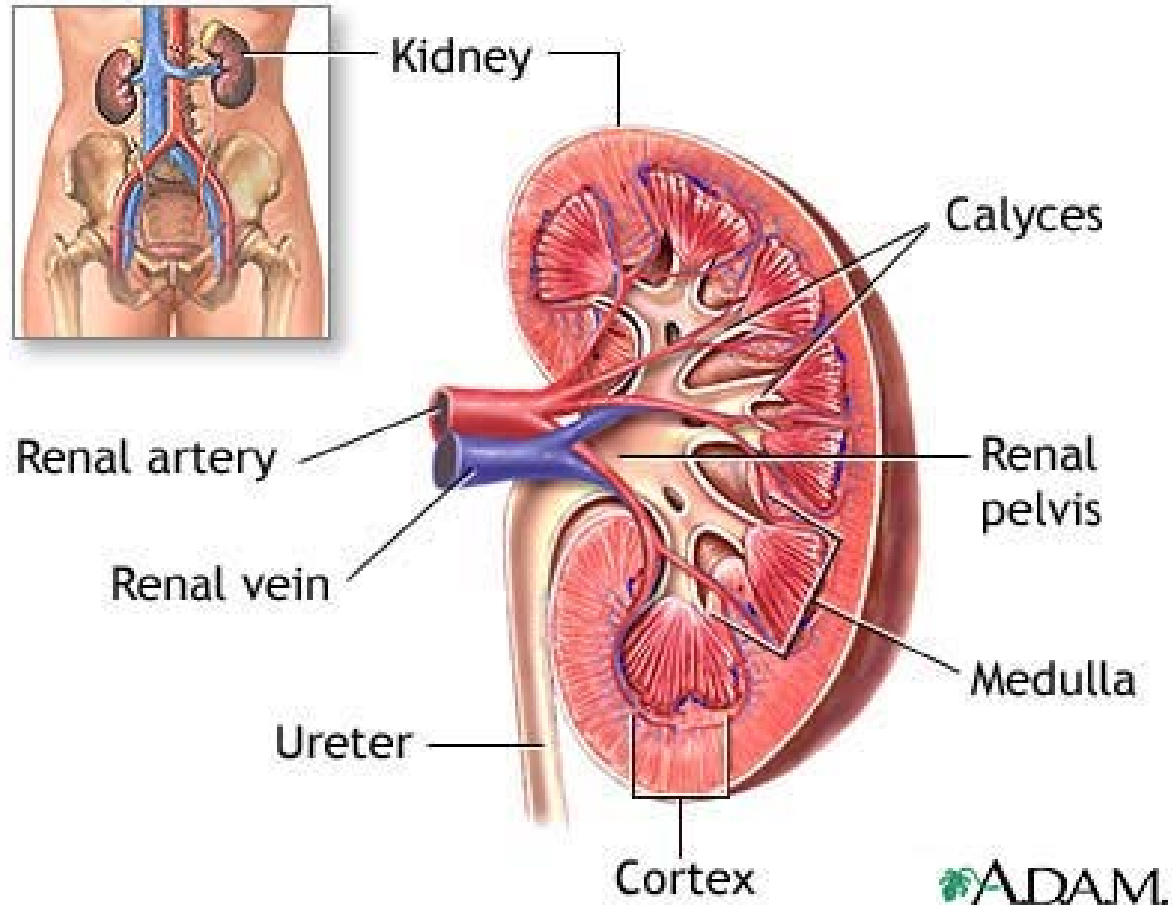
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Illustrations

Kidney Anatomy (Includes Renal Pelvis)



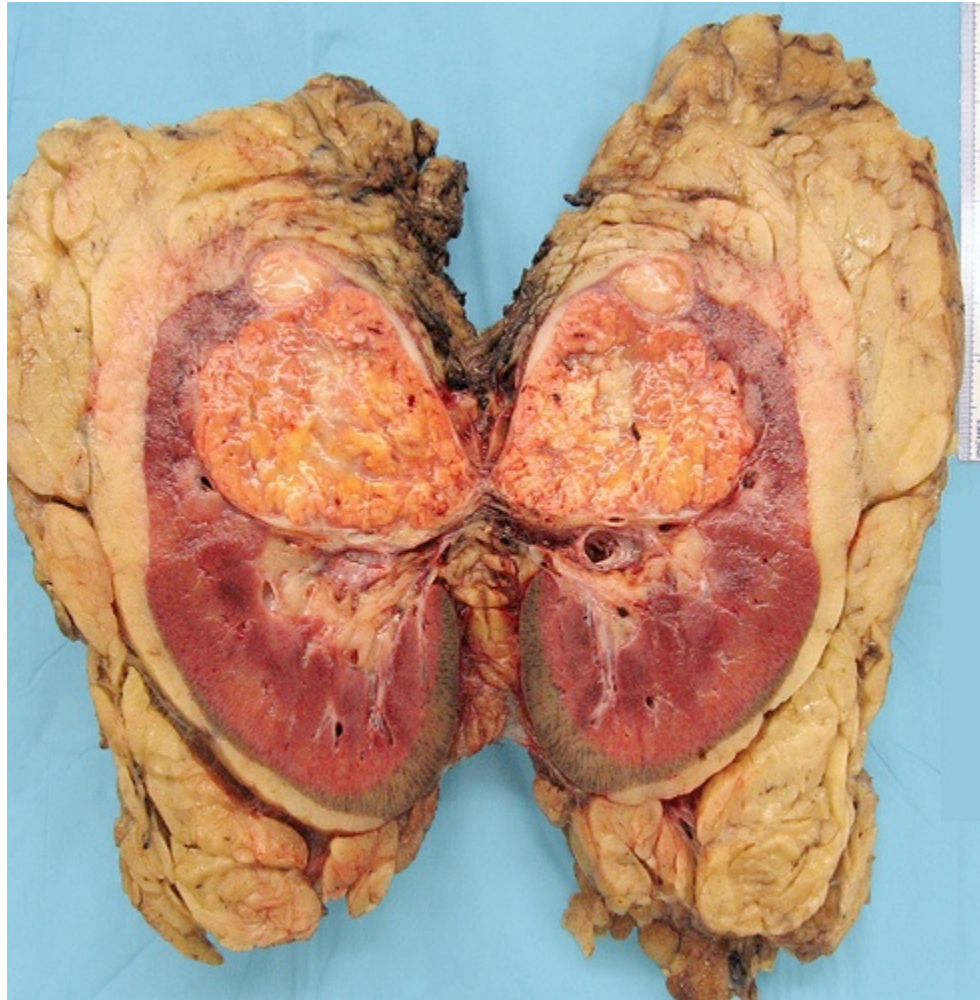
A.D.A.M. illustration used with licensed permission. All rights reserved.

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Pathology Specimen Kidneys

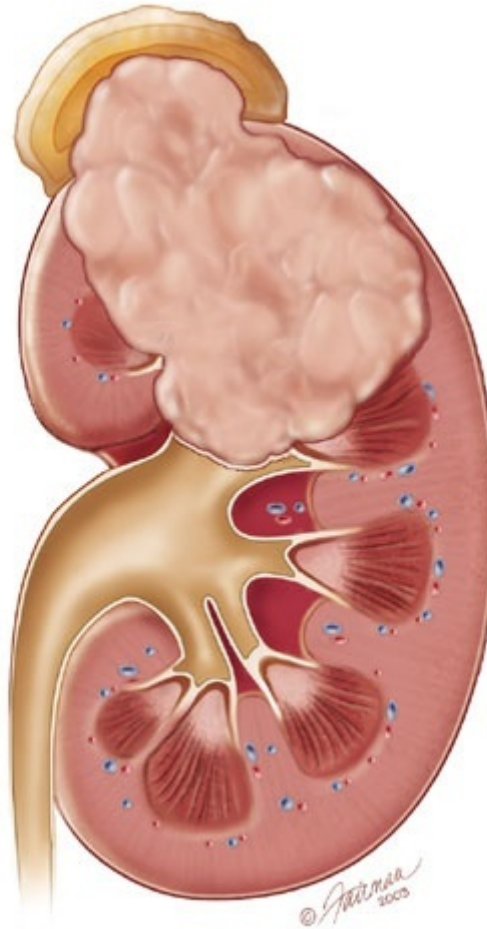


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Kidney Cancer



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

- Adrenal gland
- Bones
- Bowel
- Brain
- Discontinuous nodules in surrounding tissue
- Regional and distant lymph nodes as identified in Summary Staging Manual
- Liver
- Lung

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

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

Unknown If Single or Multiple Tumors

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

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

Note 2: Examples of cases with minimal information include

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

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

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

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Single Tumor

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

Note 1: A single tumor is always a single primary.

Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.

Note 3: The tumor may have in situ and invasive components.

Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Tumor.

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

Multiple Tumors

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

Rule M3 Abstract **multiple primaries**ⁱⁱ when **multiple tumors** are present in sites with ICD-O **site** codes that **differ** at the second (C**X**xx), third (Cx**X**x) and/or fourth characters (Cxx**X**).

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

Rule M4 Abstract a **single primary**ⁱ when there are **bilateral nephroblastomas** (previously called Wilms tumors).

Note: Timing is irrelevant; the tumors may occur simultaneously OR the contralateral tumor may be diagnosed later (no time limit).

Rule M5 Abstract **multiple primaries**ⁱⁱ when there are tumors in **both** the **right kidney** and in the **left kidney**. There may be:

- A single tumor in each kidney
- A single tumor in one kidney and multiple tumors in the contralateral kidney
- Multiple tumors in both kidneys

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

Note 2: **ONLY** abstract a single primary when **pathology** proves the tumor(s) in one kidney is/are **metastatic** from the other kidney.

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Rule M6 Abstract **multiple primaries**ⁱⁱ when the patient has a subsequent tumor after being clinically disease-free for greater than three years after the original diagnosis or last recurrence.

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

- Scans are NED
- Urine cytology is negative
- All other work-up is NED

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

Note 3: When it is unknown/not documented whether the patient had a recurrence, **default** to **date of diagnosis** to compute the time interval.

Note 4: The physician may state this is a **recurrence**, meaning the patient had a previous kidney tumor and now has another kidney tumor. **Follow the rules**; do not attempt to interpret the physician’s statement.

Note 5: The location and histology of the subsequent tumor is irrelevant. Kidney tumors that occur more than 3 years apart are always multiple primaries.

Rule M7 Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3, [Table 1](#) in the Equivalent Terms and Definitions.

Note: 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**ⁱ when synchronous, separate/non-contiguous tumors are on the **same row** in [Table 1](#) in the Equivalent Terms and Definitions. Tumors must be in the same kidney.

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **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)

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- Rule M9** Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are on **different rows** in [Table 1](#) in the Equivalent Terms and Definitions.
Note: Each row in the table is a **distinctly different** histology.
- Rule M10** Abstract a **single primary**ⁱ when an **in situ** tumor is diagnosed **after** an **invasive** tumor **AND** tumors occur in the same kidney.
Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: The tumors **may** be a **NOS** and a **subtype/variant** of that NOS. See [Table 1](#) in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.
Note 3: Once the patient has an invasive tumor, the **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.
- Rule M11** Abstract a **single primary**ⁱ (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60 days after an in situ** tumor in the same kidney.
Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS.
Note 3: When the case has been abstracted, **change behavior** code on original abstract from /2 to /3.
Note 4: Do **not** change **date of diagnosis**.
Note 5: If the case has already been submitted to the central registry, **report** all changes.
Note 6: The physician may **stage both** tumors because staging and determining multiple primaries are done for different reasons. Staging determines which treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).
Note 7: See the **COC** and **SEER** manuals for **instructions** on coding **other data items** such as Date of Diagnosis, Accession Year and Sequence Number.
- Rule M12** Abstract **multiple primaries**ⁱⁱ when an **invasive** tumor occurs **more than 60 days** after an **in situ** tumor.
Note 1: The rules are **hierarchical**. Only use this rule when none of the previous rules apply.
Note 2: Abstract **both** the invasive and in situ tumors.
Note 3: Abstract as multiple primaries even if **physician states** the invasive tumor is disease **recurrence** or **progression**.

Kidney Multiple Primary Rules

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Rule M13 Abstract a **single primary**ⁱ when there are multiple tumors that **do not meet any** of the **above criteria**.

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

Example 1: Patient presents in 2018 with renal cell carcinoma in the right kidney. Patient has a history of a previous renal cell carcinoma in the right kidney diagnosed in 2016. This is a single primary because it is the same primary site and the same histology.

Example 2: Patient presents in 2020 with a clear cell renal cell carcinoma 8310/3 in the left kidney. The patient was diagnosed with renal cell carcinoma 8312/3 in 2018. This is a single primary because it is the same primary site and a NOS and subtype/variant of that NOS.

This is the end of instructions for Multiple Tumors.

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

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

Kidney Histology Rules
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Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed prior to **neoadjuvant treatment**.
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)

Kidney Histology Rules

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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.

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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.

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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.

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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
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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

Jump to [Equivalent Terms and Definitions](#)

Jump to [Multiple Primary Rules](#)

Solid Tumor Rules
Updated November 2020

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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

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(two distinctly different histologies/different rows), code the histology from the most representative specimen (the greater amount of tumor).”

2. **New and changed** ICD-O histology codes have been added to [Table 3](#) and are identified by an asterisk. Some of those changes include:
 - A. **In situ** and **minimally invasive terms** and codes
 - B. **Terms** assigned a **new histology** code
 - C. **Histology codes** assigned a **different preferred term** (18 codes with new preferred terms)
3. The following new adenocarcinoma terms and codes have been added. The new terms and codes are **for lung only**. See [notes](#) in Table 3.
 - A. Mucinous carcinoma/adenocarcinoma
 - **8253/3** when
 - Behavior unknown/not documented (use staging form to determine behavior when available)
 - Invasive
 - **8257/3** when
 - Microinvasive
 - Minimally invasive
 - **8253/2** when
 - Preinvasive
 - In situ

Note: Previously, only **invasive /3** codes were available for mucinous adenocarcinoma of the lung. It has been recognized that not all lung cancers are invasive /3 so new codes were implemented.

- B. Non-mucinous carcinoma/adenocarcinoma
 - **8256/3** when
 - Microinvasive
 - Minimally invasive
 - **8250/2** when
 - Preinvasive
 - In situ

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C. Adenocarcinomas (CAP Terminology)

Adenocarcinoma, acinar predominant 8551

- Adenocarcinoma, lepidic predominant 8250
- Adenocarcinoma, micropapillary predominant 8265
- Adenocarcinoma, papillary predominant 8260
- Adenocarcinoma, solid predominant 8230

Equivalent or Equal Terms

These terms can be used interchangeably:

- Adenocarcinoma; carcinoma
- And; with
Note: “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Non-small cell carcinoma **8046**; a broad category which includes all histologies in Table 3 **except** for small cell carcinoma/neuroendocrine tumors (NET Tumors) **8041** and all subtypes
- Simultaneous; existing at the same time; concurrent; prior to first course treatment
- Site; topography
- Squamous cell carcinoma; SCC; epidermoid carcinoma
- Tumor; mass; tumor mass; lesion; neoplasm; nodule
 - The terms tumor, mass, tumor mass, lesion, neoplasm and nodule are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms **unless** there is a **physician’s statement** that the term is **malignant/cancer**
 - These terms are used **ONLY** to **determine multiple primaries**
 - **Do not** use these terms for **casefinding** or **determining reportability**
- Type; subtype; variant

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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**

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Table 1: Coding Primary Site

1. The mainstem bronchus **starts** at the **trachea** and extends only a few centimeters into the lung where it connects with the secondary bronchus and divides into **secondary bronchi**.
 - A. Each lobe of the lung has **secondary bronchi**
 - i. The **right** lung has **3 secondary bronchi**, one in each of the three lobes: upper; middle, and lower
 - ii. The **left** lung has **2 secondary bronchi**, one in each of the two lobes: upper and lower
 - B. Code to **mainstem bronchus** C340 when it is **specifically stated** in the operative report and/or documented by a physician.
 - C. When **only called bronchus**, code to the lobe in which the bronchial tumor is **located**
2. See the graphic in this document with the endnote “End of Mainstem Bronchus; Start of Terminal/Secondary Bronchus”
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

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Terminology	Laterality	Site Term and Code
Bronchus intermedius Carina Hilus of lung Perihilar	Bilateral	Mainstem bronchus C340 <i>Note: Bronchus intermedius is the portion of the right mainstem bronchus between the upper lobar bronchus and the origin of the middle and lower lobar bronchi</i>
Lingula of lung	Left	Upper lobe C341
Apex Apex of lung Lung apex Pancoast tumor Superior lobar bronchus Upper lobe bronchi	Bilateral	Upper lobe C341
Middle lobe Middle lobe bronchi	Right	Middle lobe C342
Base of lung Lower lobar bronchus Lower lobe Lower lobe bronchi Lower lobe segmental bronchi	Bilateral	Lower lobe C343
Overlapping lesion of lung	Bilateral	Overlapping lesion of lung C348 <i>Note: One lesion/tumor which overlaps two or more lobes</i>

Table continues on next page

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Terminology	Laterality	Site Term and Code
Bronchus NOS Bronchogenic Extending up to the hilum Extending down to the hilar region Lung NOS Pulmonary NOS Suprahilar NOS	Bilateral	Lung NOS C349 <i>Note:</i> Includes <ul style="list-style-type: none"> • Multiple tumors in different lobes of ipsilateral lung OR • Multiple tumors in ipsilateral lung; unknown if same lobe or different lobe OR • Tumor in bronchus, unknown if mainstem or lobar bronchus OR • Tumor present, unknown which lobe
Lobar bronchi NOS Lobar bronchus NOS	Bilateral	Code the lobe in which the lobar bronchus tumor is present C34__ <i>Note:</i> When lobe of origin is not documented/unknown , code to lung NOS C349

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Table 2: Combination/Mixed Histology Codes

Instructions:

1. Compare the **terms** in the **diagnosis** (pathology, cytology, radiographic, clinical) to the terms in **Column 1**.
2. When the terms **match**, use the **combination code** listed in **Column 2**.
3. The **last row** in the table is a “**last resort**” code: adenocarcinoma mixed subtypes 8255.

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

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

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

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

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

Column 1 lists the **required terms for the combination code**.

Column 2 lists the **combination term and code** for histologies **in Column 1**.

Table begins on next page.

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Required Terms	Combination Histologies and Code
Adenocarcinoma NOS <p style="text-align: center;">AND</p> Squamous cell carcinoma NOS <i>Note:</i> Diagnosis must be adenocarcinoma NOS and squamous cell carcinoma NOS, NOT any of the subtypes/variants of adenocarcinoma or squamous cell carcinoma	Adenosquamous carcinoma 8560
Giant cell carcinoma <p style="text-align: center;">AND</p> Spindle cell carcinoma <i>Note:</i> Sarcomatoid carcinoma is not in the histology table because sarcomatoid tumors primarily originate in the mediastinum. The combination code is added for the rare occasion when a tumor occurs within the lung.	Sarcomatoid carcinoma 8033 <i>Note:</i> Both giant cell carcinoma and spindle cell carcinoma are components of sarcomatoid carcinoma. The most accurate code for a combination of giant cell and spindle cell carcinoma is sarcomatoid carcinoma
Epithelial carcinoma <p style="text-align: center;">AND</p> Myoepithelial carcinoma	Epithelial-myoepithelial carcinoma 8562
Mucinous carcinoma, invasive <p style="text-align: center;">AND</p> Non-mucinous carcinoma, invasive	Mixed invasive mucinous and non-mucinous carcinoma 8254/3*

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Required Terms	Combination Histologies and Code
<p>Small cell carcinoma/neuroendocrine tumor (NET) <i>Note: Includes subtypes/variants</i> of small cell/neuroendocrine tumor. See Table 3 for subtypes/variants.</p> <p style="text-align: center;">AND</p> <p>At least one of the following:</p> <ul style="list-style-type: none"> • 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 	<p>Combined small cell carcinoma 8045</p>
<p>Squamous cell carcinoma (epidermoid carcinoma)</p> <p style="text-align: center;">AND</p> <p>Large cell non-keratinizing squamous cell carcinoma</p> <p><i>Note: Squamous cell carcinoma and epidermoid carcinoma are synonyms</i></p>	<p>Squamous cell carcinoma, large cell, nonkeratinizing 8072</p>
<p>Squamous cell carcinoma (epidermoid carcinoma)</p> <p style="text-align: center;">AND</p> <p>Small cell nonkeratinizing squamous cell carcinoma</p> <p><i>Note: Squamous cell carcinoma and epidermoid carcinoma are synonyms</i></p>	<p>Squamous cell carcinoma, small cell, nonkeratinizing 8073</p>

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Required Terms	Combination Histologies and Code
Squamous cell carcinoma, keratinizing <p style="text-align: center;">AND</p> Squamous cell carcinoma, non-keratinizing	Squamous cell carcinoma, NOS 8070
Squamous cell (epidermoid) carcinoma <p style="text-align: center;">AND</p> One or both of the following: <ul style="list-style-type: none"> • Sarcomatoid carcinoma • Spindle cell carcinoma <p><i>Note 1:</i> Does not include subtypes/variants of squamous cell. See Table 3 for subtypes/variants.</p> <p><i>Note 2:</i> Squamous cell carcinoma and epidermoid carcinoma are synonyms.</p>	Squamous cell carcinoma, sarcomatoid 8074 Squamous cell carcinoma, spindle cell 8074

Table continues on next page

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Required Terms	Combination Histologies and Code
<p>Diagnosis must be a single tumor which meets one of the following two criteria:</p> <ol style="list-style-type: none"> 1. <u>At least two</u> of the subtypes/variants of adenocarcinoma AND <u>percentages of each type are unknown/not stated</u> <ul style="list-style-type: none"> • Acinar adenocarcinoma • Clear cell adenocarcinoma • Lepidic adenocarcinoma <i>Note:</i> Lepidic adenocarcinoma may or may not have mucinous components. • Micropapillary adenocarcinoma • Papillary adenocarcinoma • Solid adenocarcinoma • Well-differentiated fetal adenocarcinoma <i>Note:</i> 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. 	<p>Adenocarcinoma with mixed subtypes 8255/3 <i>Note 1:</i> 8255 is a “last resort” code. <i>Note 2:</i> See the Histology Rules to determine when it is appropriate to use this code for combination histologies other than adenocarcinoma subtypes/variants. <i>Note 3:</i> 8255 does not apply to squamous cell carcinoma, NOS and/or subtype/variants of SCC.</p>

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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

Lung Equivalent Terms and Definitions
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Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
<p>Adenocarcinoma 8140</p> <p>Note 1: Mucinous adenocarcinoma for lung only is coded as follows:</p> <ul style="list-style-type: none"> • 8253/3* when <ul style="list-style-type: none"> ○ Behavior unknown/not documented (use staging form to determine behavior when available) ○ Invasive • 8257/3* when <ul style="list-style-type: none"> ○ Microinvasive ○ Minimally invasive • 8253/2* when <ul style="list-style-type: none"> ○ Preinvasive ○ In situ <p>Note 2: Non-mucinous adenocarcinoma for lung only is coded as follows:</p> <ul style="list-style-type: none"> • 8256/3* when <ul style="list-style-type: none"> ○ Microinvasive ○ Minimally invasive • 8250/2* when <ul style="list-style-type: none"> ○ Preinvasive ○ In situ 	<p>Adenocarcinoma NOS</p> <p>Adenocarcinoma in situ 8140/2</p> <p>Adenocarcinoma invasive 8140/3</p> <p>Adenocarcinoma, non-mucinous, NOS</p>	<p>Acinar adenocarcinoma/adenocarcinoma, acinar predominant (for lung only) 8551*</p> <p>Adenoid cystic/adenocystic carcinoma 8200</p> <p>Colloid adenocarcinoma 8480</p> <p>Fetal adenocarcinoma 8333</p> <p>Lepidic adenocarcinoma/adenocarcinoma, lepidic predominant 8250/3*</p> <p>Mucinous carcinoma/adenocarcinoma (for lung only)</p> <ul style="list-style-type: none"> in situ 8253/2* invasive 8253/3* minimally invasive 8257/3* microinvasive 8257/3* preinvasive 8253/2* <p>Micropapillary adenocarcinoma/adenocarcinoma, micropapillary predominant 8265</p> <p>Mixed invasive mucinous and non-mucinous adenocarcinoma 8254*</p> <p>Non-mucinous adenocarcinoma (for lung only)</p> <ul style="list-style-type: none"> in situ 8250/2* microinvasive 8256/3* minimally invasive 8256/3* preinvasive 8250/2* <p>Papillary adenocarcinoma/adenocarcinoma, papillary predominant 8260</p> <p>Pulmonary intestinal-type adenocarcinoma/enteric adenocarcinoma 8144</p> <p>Solid adenocarcinoma/adenocarcinoma, solid predominant 8230</p>

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Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Adenosquamous carcinoma 8560		
Epithelial-myoepithelial carcinoma 8562 <i>Note:</i> Adenomyoepithelioma, epithelial/myoepithelial tumor of unproven malignant potential were thought to be adenomas (not reportable) prior to 2018. These histologies are now designated as low-grade carcinomas based on lymph node metastasis, local invasion, and aggressiveness	Adenomyoepithelioma* Epimyoeplithelial carcinoma Epithelial-myoepithelial tumor of unproven malignant potential* Malignant mixed tumor comprising epithelial and myoepithelial cells Pneumocytic adenomyoepithelioma*	
Epithelioid hemangioepithelioma 9133		
Giant cell carcinoma 8031		
Intrapulmonary thymoma (arising within lung) 8580/3 <i>Note:</i> <u>Intrapulmonary</u> thymoma is always malignant /3.		

Table continues on next page

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Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
<p>Large cell carcinoma 8012</p> <p><i>Note 1:</i> A diagnosis of large cell carcinoma is usually followed by further diagnostic testing to identify the subtype/variant.</p> <p><i>Note 2:</i> 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.</p> <p><i>Note 3:</i> Large cell carcinoma with neuroendocrine (NE) differentiation lacks NE morphology and is coded as large cell carcinoma, <u>not</u> large cell neuroendocrine carcinoma.</p>	<p>Large cell anaplastic carcinoma</p> <p>Large cell carcinoma NOS</p> <p>Large cell carcinoma with no additional stains (subtype/variant – no ICD-O code)</p> <p>Large cell carcinoma with null immunohistochemical features (subtype/variant – no ICD-O code)</p> <p>Large cell carcinoma with unclear immunohistochemical features (subtype/variant – no ICD-O code)</p> <p>Large cell undifferentiated carcinoma</p>	
Lymphoepithelioma-like carcinoma 8082		
Melanoma 8720		
<p>Mucoepidermoid carcinoma 8430</p> <p><i>Note:</i> Mucoepidermoid tumor is listed as a synonym of mucoepidermoid carcinoma in WHO</p>	Mucoepidermoid tumor	
Myoepithelial carcinoma 8982		

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Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
NUT carcinoma 8023/3* NUT: nuclear protein in tests NUT/M1 gene rearrangement	Aggressive t(15:19) positive carcinoma BET-rearranged carcinoma Carcinoma with t(15:19) translocation Midline carcinoma of children and young adults with NUT rearrangement Midline lethal carcinoma NUT midline carcinoma	
PEComa malignant 8714/3 <i>Note:</i> Tumor displays perivascular epithelioid (PEC) differentiation		
Pleomorphic carcinoma 8022 <i>Note 1:</i> The definition of pleomorphic carcinoma is that it is a subtype of sarcomatoid carcinoma. It has at least 10% spindle or giant cells <i>Note 2:</i> Pleomorphic carcinoma has components of adenocarcinoma and/or large cell carcinoma, also squamous carcinoma		

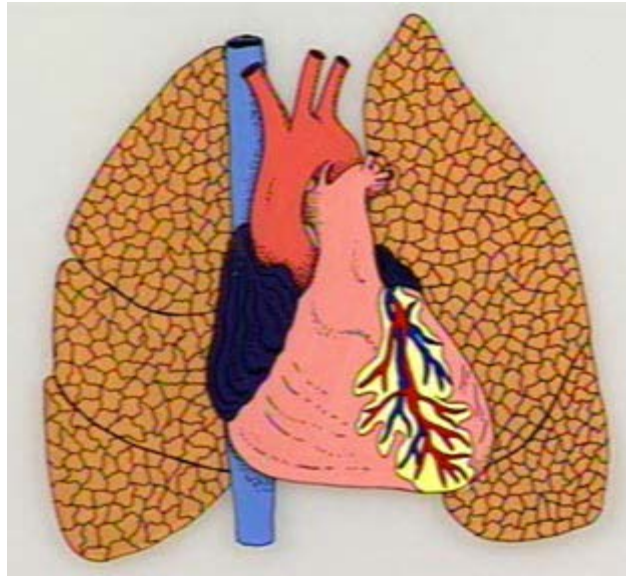
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Specific or NOS Histology Term and Code	Synonym of Specific or NOS	Subtype/variant of NOS and Code
Sarcoma NOS 8800/3		Biphasic synovial sarcoma 9043/3 Epithelioid cell synovial sarcoma 9042/3 Pulmonary artery intimal sarcoma/low-grade malignant myxoid endobronchial tumor 9137/3 Pulmonary myxoid sarcoma with EWSR1 - CREB1 translocation 8842/3 Spindle cell synovial sarcoma 9041/3 Synovial sarcoma 9040/3
Small cell carcinoma 8041/3 <i>Note 1:</i> This row applies to neuroendocrine tumors (NET). <i>Note 2:</i> Large cell carcinoma with neuroendocrine differentiation lacks NE morphology and is coded as large cell carcinoma, not large cell neuroendocrine carcinoma.	Reserve cell carcinoma Round cell carcinoma SCLC Small cell carcinoma NOS Small cell neuroendocrine carcinoma	Atypical carcinoid 8249/3 Combined small cell carcinoma 8045/3 Large cell neuroendocrine carcinoma/combined large cell neuroendocrine carcinoma 8013/3 Typical carcinoid 8240/3 Neuroendocrine carcinoma, NOS Well-differentiated neuroendocrine carcinoma
Spindle cell carcinoma 8032		
Squamous cell carcinoma 8070	Epidermoid carcinoma Epidermoid carcinoma NOS Squamous carcinoma Squamous cell carcinoma NOS Squamous cell epithelioma Squamous cell carcinoma in situ 8070/2	Basaloid carcinoma/basaloid squamous cell carcinoma 8083 Keratinizing squamous cell carcinoma 8071 Non-keratinizing carcinoma 8072

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

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

Illustrations



Used with permission

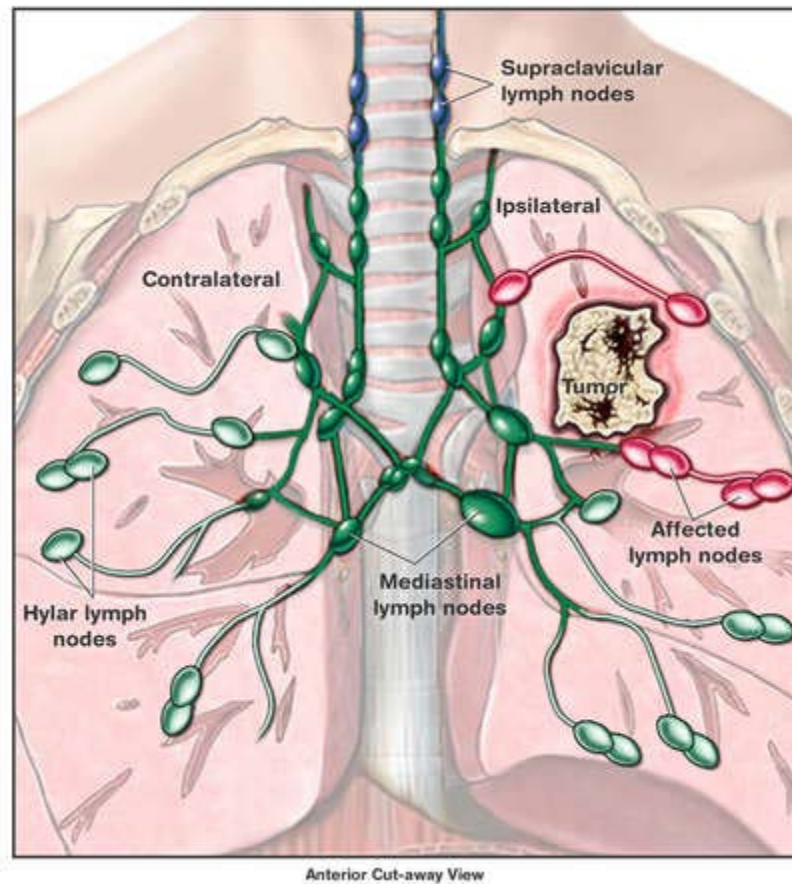
Lung Equivalent Terms and Definitions

C340-C343, C348, C349

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

Mediastinum

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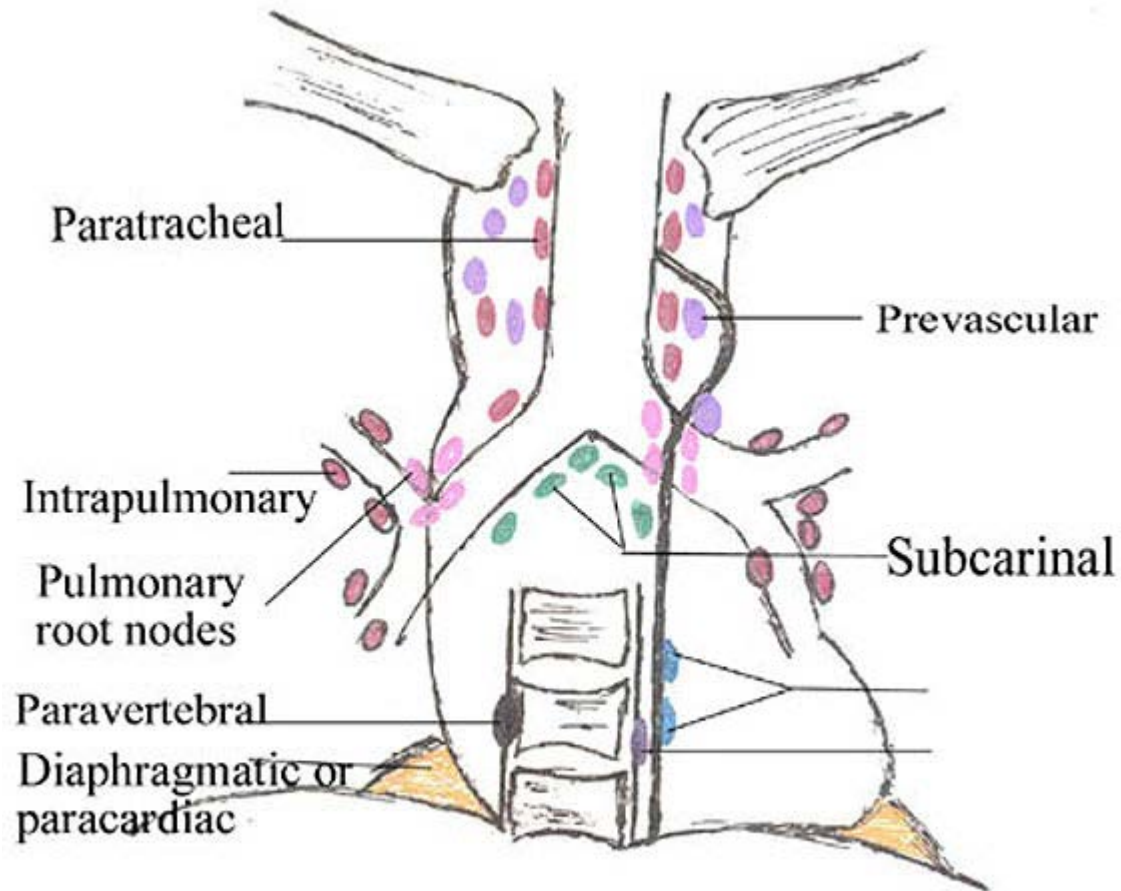
Lung Equivalent Terms and Definitions

C340-C343, C348, C349

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

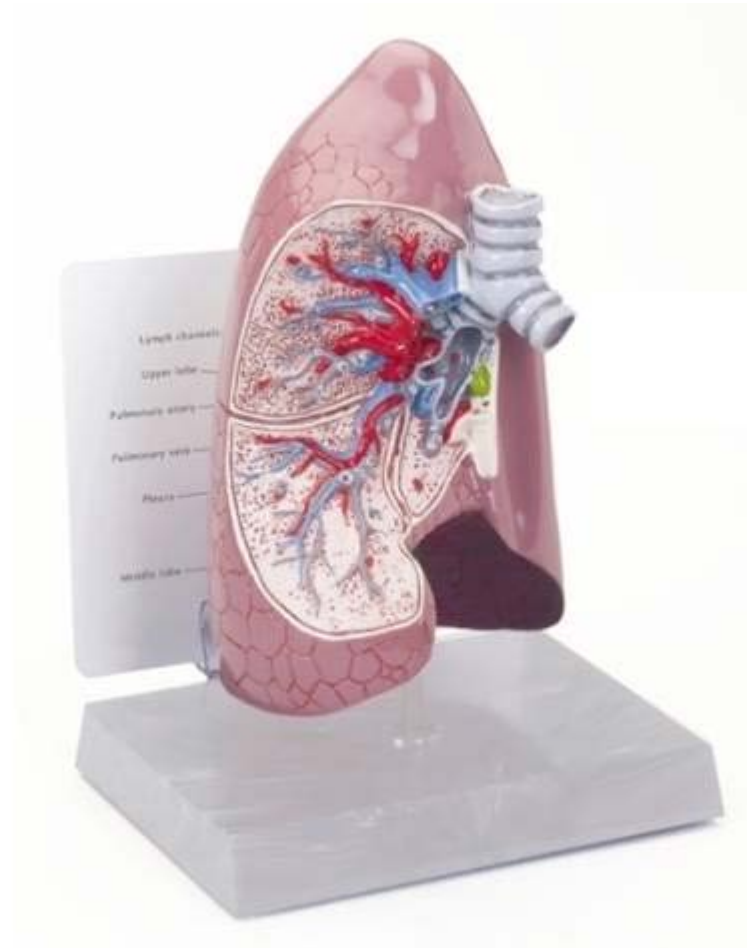
Lymph Nodes Lung

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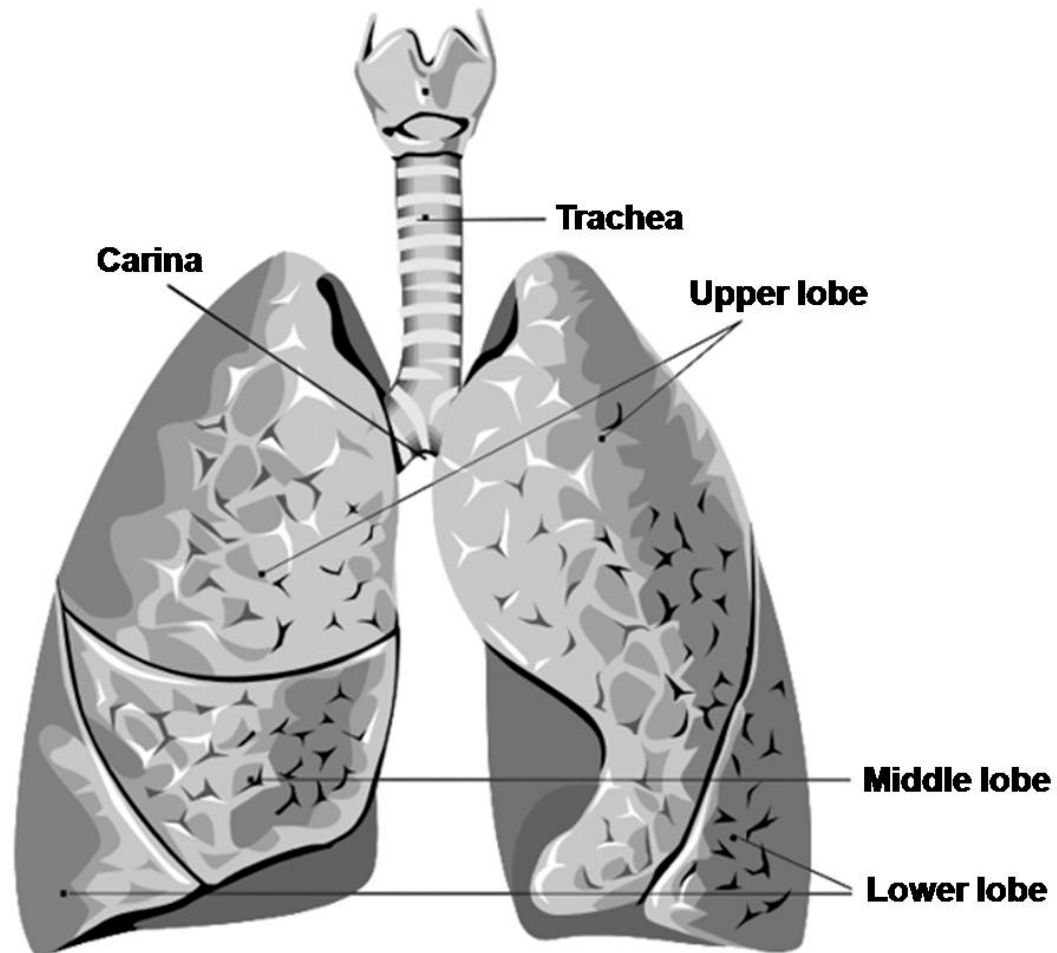
Lung Equivalent Terms and Definitions
C340-C343, C348, C349
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Gross Anatomy of Lung
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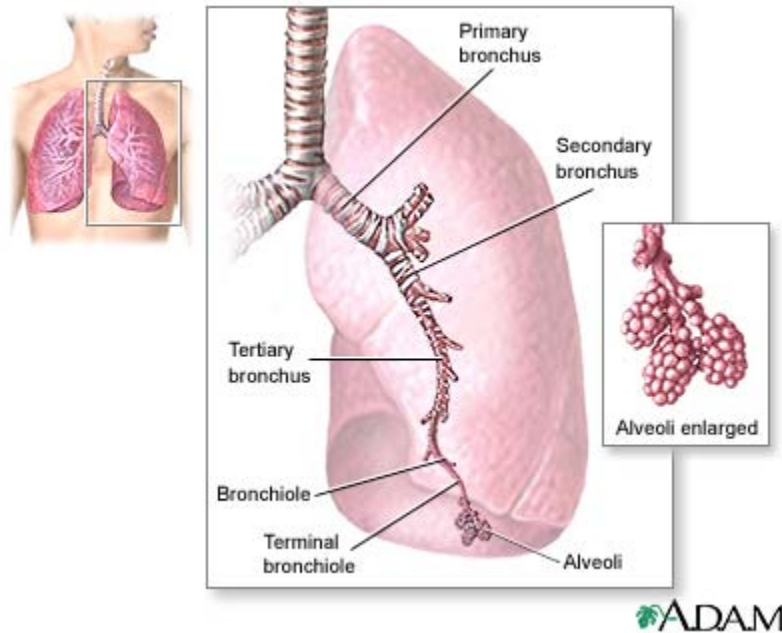
Lung Equivalent Terms and Definitions

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**ⁱ when it is not possible to determine if there is a **single** tumor or **multiple** tumors.

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

Note 2: Examples of cases with minimal information include

- Death certificate only (DCO)
- Cases for which information is limited to pathology report only
 - o Outpatient biopsy with no follow-up information available
 - o 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

ⁱPrepare 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**ⁱⁱ when there are **separate, non-contiguous** tumors in sites with ICD-O **site** codes that differ at the second **CX**xx and/or third character **Cx****X**x.

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

Rule M4 Abstract **multiple primaries**ⁱⁱ when the patient has a subsequent tumor after being **clinically disease-free** for greater than **three years** after the original diagnosis or last recurrence.

Note 1: **Clinically** disease-free means that there was **no 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**ⁱⁱ when there is **at least one** tumor that is **small cell carcinoma 8041** or any small cell subtypes/variants and another tumor that is **non-small cell carcinoma 8046** or any non-small cell carcinoma subtypes/variants.

Note 1: Small cell carcinoma and non-small cell carcinoma are the two major classifications/divisions for lung cancer.

- See [Table 3](#) in Equivalent Terms and Definitions for terms and codes for small cell carcinoma and all of the subtypes/variants
- With the exception of small cell/neuroendocrine carcinoma and sarcomas, **all other histologies** listed in [Table 3](#) in Equivalent Terms and Definitions are **non-small cell carcinoma**

Note 2: It is **irrelevant** whether the tumors are in the **ipsilateral** (same) lung or are **bilateral** (both lungs).

Rule M6 Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3, [Table 3](#) in the Equivalent Terms and Definitions. Timing is irrelevant.

Note: 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**ⁱ when synchronous, separate/non-contiguous tumors **in the same lung** are on **the same row** in [Table 3](#) in the Equivalent Terms and Definitions.

Note 1: Tumors must be **in the same lung**.

Note 2: The same row means the tumors are:

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

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

Rule M8 Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are:

- On different rows in [Table 3](#) in the Equivalent Terms and Definitions
- A combination code in [Table 2](#) and a code from [Table 3](#)

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

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

Example 1: In 2018, the patient has non-mucinous adenocarcinoma **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**ⁱ when there are **simultaneous multiple** tumors:

- In **both** lungs (multiple in right and multiple in left) **OR**
- In the **same** lung **OR**
- **Single** tumor in one lung; **multiple** tumors in **contralateral** lung

Note 1: Tumors may be combinations of:

- In situ and invasive **OR**
- NOS and subtype/variant (See [Table 3](#) in the Equivalent Terms and Definitions)

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
 - o **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**ⁱ when an **in situ** tumor is diagnosed **after** an **invasive** tumor **AND** tumors occur in the same lung.

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

Note 2: The tumors may be a **NOS** and a **subtype/variant** of that NOS. See [Table 3](#) in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

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

Rule M11 Abstract **multiple primaries**ⁱⁱ when there is a **single** tumor in **each lung** (one tumor in the right lung and one tumor in the left lung).

Note 1: The only **exception** is when there is **proof** that one tumor is **metastatic**. Proof is any one of the following:

- Tissue from both tumors is compared and the pathologic diagnoses definitively says one tumor is metastatic
- Attending physician, oncologist, or pulmonologist state unequivocally that the tumor in the contralateral lung is metastatic
 - o Unequivocal means that no words such as “probably possibly, most likely, etc.” are used in the statement. Terms which are on the “ambiguous terms” list make the statement equivocal (cannot be used to prove metastases)

Note 2: Lung **metastases usually** present as multiple tumors/masses. A single tumor in each lung is unlikely to be a single primary (e.g. metastatic to the contralateral lung).

Note 3: The term “**bilateral**” is **not** a synonym for a **single primary**. It is simply a statement that there are tumors in both lungs.

Note 4: This rule is based on **long-term epidemiologic studies** of multiple primaries. The specialty medical experts (SME) and the CoC site physician teams reviewed and approved these rules. Many of the CoC site team physicians were also authors, co-authors, or editors of the AJCC Staging Manual.

Note 5: Lymph node involvement is recorded in staging criteria.

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

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

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

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

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

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

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

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

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

Rule M13 Abstract **multiple primaries**ⁱⁱ when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor in the same lung.

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

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

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

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

Rule M14 Abstract a **single primary**ⁱ when none of the previous rules apply.

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

This is the end of instructions for Multiple Tumors

ⁱPrepare 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.

ⁱⁱPrepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

**Lung Histology Rules
C340-C343, C348, C349**

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

Note: WHO 4th Ed Tumors of Lung: in 2011 has a new classification of adenocarcinoma which is a significant changes from the 2004 WHO classification. One of the major changes is discontinuing usage of the term bronchioloalveolar carcinoma (BAC) beginning with cases diagnosed 1/1/2018 and forward. The preferred term for BAC is now mucinous adenocarcinoma **8253**.

Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed prior to neoadjuvant treatment.
Note 1: Histology changes 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**, 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.

**Lung Histology Rules
C340-C343, C348, C349**

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

3. Code the specific histology described by **ambiguous terminology** (list follows) **ONLY** when A or B is true:
- A. The only diagnosis available is **one histology** term described by ambiguous terminology
- CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
 - Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented
- Example:* Outpatient biopsy says probably squamous cell carcinoma. The case is accessioned (entered into the database) as required by both SEER and COC. No further information is available. Code the histology squamous cell carcinoma. The case meets the criteria in **#3A**.
- B. There is a **NOS histology and a more specific** (subtype/variant) described by ambiguous terminology
- Specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.) **OR**
 - Patient is receiving treatment based on the specific histology described by ambiguous term
- Example:* The pathology diagnosis is NSCLC consistent with adenocarcinoma. The oncology consult says the patient has adenocarcinoma of the right lung. This is clinical confirmation of the diagnosis, code adenocarcinoma. The case meets the criteria in **bullet 1**.
- Example:* The pathology diagnosis is NSCLC consistent with squamous cell carcinoma. The treatment plan says the patient will receive treatment for squamous cell carcinoma. Treatment plan confirms squamous cell carcinoma; code squamous cell carcinoma. The case meets the criteria in **bullet 2**.

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

List of Ambiguous Terminology

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

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

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/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: 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.

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

Rule H4 Code the histology when only **one histology** is present.

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

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

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

Note 4: This includes coding non-small cell carcinoma when it is the only diagnosis available.

Rule H5 Code the **invasive** histology **when in situ** and **invasive** histologies are present.

Note 1: Histologies may be **NOS** and a **subtype/variant**.

Note 2: When the NOS is invasive and the subtype/variant is situ, code the NOS (invasive).

Example: The histologies are mucinous adenocarcinoma in situ **8253/2** and invasive adenocarcinoma NOS **8140/3**. Code the invasive histology: adenocarcinoma **8140/3**.

Rule H6 Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS, such as the following:

- Adenocarcinoma **8140** and a subtype/variant of adenocarcinoma
- Mucinous adenocarcinoma and a subtype/variant of mucinous adenocarcinoma
- Non-small cell carcinoma **8046** and a subtype/variant of non-small cell carcinoma
- Sarcoma **8800** and a subtype/variant of sarcoma
- Small cell neuroendocrine tumors/NET **8041** and a subtype/variant of small cell neuroendocrine tumor/NET
- Squamous cell carcinoma **8070** and a subtype/variant of squamous cell carcinoma

Note: See [Table 3](#) in the Equivalent Terms and Definitions to find NOS and subtypes/variants.

**Lung Histology Rules
C340-C343, C348, C349**

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

Rule H7 Code the histology that comprises the **greatest percentage** of tumor when two or more of the following histologies are present:

- Acinar adenocarcinoma / Adenocarcinoma, acinar predominant **8551**
- Lepidic adenocarcinoma / Adenocarcinoma, lepidic predominant **8250**
- Micropapillary adenocarcinoma / Adenocarcinoma, micropapillary predominant **8265**
- Papillary adenocarcinoma / Adenocarcinoma, papillary predominant **8260**
- Solid adenocarcinoma / Adenocarcinoma, solid predominant **8230**

Note 1: The rules are hierarchical, so the tumors are **NOT** a NOS and subtype/variant.

Note 2: If the percentages are unknown/not documented, 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.

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.

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Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
Note: “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Cerebrospinal fluid; CSF
- Dura; meninges
- Extradural; not within the meninges; within the cranium; within the skull but not within the cerebral meninges
- Extramedullary; outside medulla oblongata C700
- Infratentorial; below the tentorium cerebelli; either cerebellum or brainstem
- Intracranial; within the skull, within the cranium
- Intradural; between layers of the cerebral meninges; C700
- Intradural-extramedullary; within the spinal canal but outside of the nerves
- Intraspinal; occurring within the spinal column especially the vertebral canal; spinal nerve roots
- Site; topography
- Supratentorial; above the tentorium cerebelli; cerebrum
 - Cerebrum contains frontal lobe, parietal lobe, occipital lobe, and temporal lobe
- Tentorium cerebelli; cerebellar tentorium
- Tumor; mass; lesion; neoplasm when
 - These terms are used ONLY to determine multiple primaries
 - Do not use these terms for casefinding or determining reportability
- Type; subtype; variant
- WHO Grade 3 and WHO Grade 4; malignant; invasive; /3

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Terms that are NOT Equivalent or Equal

This is a term that is **not equivalent**. There are no casefinding implications.

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

Changes from 2007 MPH Rules

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

1. 2016 CNS WHO presents major restructuring of the diffuse gliomas, medulloblastomas and other embryonal tumors, and incorporates new entities that are defined by both histology and molecular features, including glioblastoma, IDH-wildtype and glioblastoma, IDH-mutant; diffuse midline glioma, H3 K27M-mutant, RELA fusion-positive ependymoma, medulloblastoma, WNT-activated and medulloblastoma, SHH-activated, and embryonal tumor with multilayered rosettes, C19MC-altered. The 2016 edition has added newly recognized neoplasms and has referred to some entities, variants and patterns as “not recommended” (previously called obsolete).
 - A. It has been determined that these “not recommended” terms no longer have diagnostic and/or biological relevance. For example, gliomatosis cerebri is a term which is no longer recommended. Gliomatosis cerebri is now termed a “growth pattern” rather than a histologic type.
 - B. Terms which are not recommended are not included in the tables. When one of these terms are used, refer to the ICD-O and all updates for the correct histology code. For example, glioma NOS is an umbrella term for all gliomas and astrocytomas. Glioma NOS is not recommended because diagnostic methodology is able to determine a more specific diagnosis.
2. **Rule change:** The 2007 rules said a glioblastoma multiforme (GBM) following an astrocytic or glial tumor was a single primary (recurrence). In the 2018 Solid Tumor Rules, GBM subsequent to an astrocytic or glial tumor is a multiple primary. GBM is now being collected as a new primary so it is possible to analyze the frequency with which these tumors recur in a more aggressive form (GBM).

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3. Clarifications:

- A. The following meningiomas are reportable: intraosseous, cavernous sinus and sphenoid wing.
 - B. Multiple cerebral meningiomas are a single primary.
 - C. Multiple brain tumors (same histology) are a single primary.
 - D. Laterality is not used to determine multiple primaries.
 - E. Timing is not used to determine multiple primaries.
 - F. The brain (C710-C719) is a single primary site.
 - G. Neurofibromatosis NOS, Neurofibromatosis 1 (NF1), Neurofibromatosis 2 (NF2), and schwannomatosis are genetic syndromes and not reportable neoplasms. People with this genetic syndrome do have a high risk of developing:
 - i. Non-reportable non-malignant tumors occurring in skin and sites other than CNS AND
 - ii. Reportable malignant tumors
4. The 2016 World Health Organization (WHO) Classification of Tumors of the Central Nervous System (CNS) contains newly recognized histologies and subtypes/variants. New codes/terms are identified by asterisks (*) in **Table 3** in the Terms and Definitions.

Reportability Criteria

CNS neoplasms must meet all **three of the conditions below** to be **reported as malignant /3**:

- 1. The **behavior** must be malignant /3:
 - A. Pathology designates the behavior as malignant/invasive, /3 **OR**
 - B. The tumor is a WHO Grade 3 or 4 (See **Section 1: Table 1**)

Note 1: WHO Grade 2 tumors may be non-malignant or malignant.
Note 2: **Always** code the behavior as designated by the **pathologist**.
Note 3: **Never report** a **malignant /3** behavior code for a meningioma **based on tumor extension** to brain, skin of scalp, or other regional organs/tissue. Non-malignant CNS tumors can extend to the regional tissue and bone.
- 2. The **primary site** must be reportable (See **Section 2: Table 2**) **AND**
- 3. The **histology** must be reportable (See **Section 2: Table 3**)

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Directory of Sections and Tables

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

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

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Section 1: Behavior Code

Note: Behavior determines which set of CNS rules should be used: malignant or non-malignant.

Instructions for using source **documentation** to determine behavior are in **priority order**. Start with Instruction 1 and **STOP** when you reach the instruction that applies to the case being abstracted. For example, when the **resection pathology** specifies behavior, use Instruction 1. **Do not** code the behavior using Instructions 2-5.

Priority Order for Using Documentation to Assign Behavior

1. Pathology: Tissue from **resection**
 - A. Use the pathologist's **description** of **malignant/invasive** behavior
 - B. Cases are reportable as malignant when pathology states a WHO Grade 3 or 4
 - i. WHO Grade 2 may be either non-malignant or malignant. Use the pathologist's description of behavior (priority #1).
 - C. **Never change behavior** described by pathologist
2. Pathology: Tissue from **biopsy**
3. Cytology (usually cerebrospinal fluid)
4. Physician's documentation (no pathology report) in the following priority order:
 - A. Tumor Board
 - B. Documentation of original pathologic diagnosis and behavior
Example: Pathology report is not in the medical record. Oncology consult states biopsy was done in a different facility and cites the **original pathology** diagnosis including the **behavior**.
 - C. **Documentation** of behavior, no mention of original diagnosis
Example: Pathology report is not in the medical record. **Physician documents** the **behavior** as malignant, or WHO Grade 3 or 4, but **does not cite/mention original** pathology report as source of behavior classification.
5. **Scan**, use behavior information from radiography in the following **priority order**:
 - A. MRI
 - B. CT
 - C. PET
 - D. Angiogram

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

Table 1: WHO Grades for Select CNS Neoplasms

Note 1: CNS WHO classifications use a grading scheme that is a "malignancy scale" ranging across a wide variety of neoplasms rather than a strict histologic grading system that can be applied equally to all tumor types.

Note 2: See the SEER and COC Manuals for instructions on coding grade for CNS tumors.

Note 3: The table **does not** contain all neoplasms that may occur in the CNS.

Note 4: WHO does not provide Grades for all CNS and peripheral nerve neoplasms.

WHO Grade Definitions

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

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 I** (always non-malignant).

Column 1 contains the **histology** term.

Column 2 contains the **WHO Grade** assigned **based** on the **molecular features** of the histology.

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

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

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

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Section 2: Reportable Primary Sites and Histologies

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

Note 1: Peripheral nerves are included in the Malignant CNS and Peripheral Nerve rules because:

- **All malignant** tumors are reportable, including peripheral nerve tumors **AND**
- The Malignant CNS and Peripheral Nerve **rules** contain the **correct histologies** and coding **rules** for tumors of **peripheral nerves and meninges/dura.**

Note 2: Peripheral nerves are:

- **Extracranial**/outside the cranium **OR**
- **Extradural**/outside the spinal dura

Note 3: The following malignant **meningiomas** are reportable:

- **Intraosseous**

Note: The **dura** layer of the meninges **contacts** the **endosteum** of the bones of the skull. The primary site for intraosseous meningioma is cranial meninges C700.

- **Sphenoid wing**

Note 1: Sphenoid wing meningiomas arise in the **cranial meninges** C700 which covers the bony structure called the sphenoid wing.

Note 2: The term “sphenoid wing meningioma” is used to identify the **location** of the meningioma because sphenoid wing meningiomas may be very **invasive**, spreading to the dura of the frontal, temporal and orbital regions.

- **Cavernous sinus**

Note 1: Cavernous sinus is located between the endosteal and meningeal layers of the dura.

Note 2: There is **no ICD-O site** code for cavernous sinus because tumors do not arise in the sinus itself; the primary sites are:

- The **cranial nerves** passing through the sinus (trochlear, abducent **C725**) **OR**
- The cerebral **meninges/dura** **C700** covering the cranial nerve

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Priorities for Coding Primary Site

Note 1: Always check the **operative report(s)** which will have **information** on whether the surgery or biopsy was **intracranial** (inside the cranium/skull) or **intraspinal** (within the dura/meninges covering the spinal cord).

Note 2: Code **the specific primary site**. Use an NOS site code only when a specific site is not known.

Use the list in **hierarchical order**:

1. **Resection**

- A. Operative report(s)
- B. Pathology report(s)

2. **Biopsy**

- A. Operative report(s)
- B. Pathology report(s)

3. Resection and/or biopsy performed, but operative report(s) and pathology are not available (minimal information)

- A. Tumor Board
- B. Code from physician's documentation of **original diagnosis** from operative or pathology report OR
- C. Physician's **documentation of primary site** in the medical record

Example: The patient had a **biopsy** done at **another facility**. The operative and pathology reports are not in the medical record. The **attending documents** that the patient had a biopsy of a **cerebral meningioma** at a different facility. Code the primary site cerebral meninges C700.

4. For cases diagnosed by imaging (**no pathology/resection or biopsy**) use information from scans in the following priority order:

- A. MRI
- B. CT
- C. PET
- D. Angiogram

5. See **Table 2: Reportable Primary Sites** to confirm the primary site is reportable.

6. When the primary site is cranial nerve **OR** peripheral nerve, see **Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves** to determine whether the portion of the nerve is cranial or peripheral (different site codes).

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Reportable Primary Site Groups

The three major **groups** of reportable sites are:

1. **Intracranial** (within the skull/cranium) **AND**
2. **Spinal sites** (spinal meninges and sites within the spinal meninges)
3. **Peripheral nerves** (extracranial or extraspinal nerves).

Terms and sites for these broad categories are:

Reportable Primary Sites and their ICD-O Codes

1. Intracranial (intra means inside or within; intracranial is inside the skull).
 - A. **Cerebral/cranial dura/meninges** C700. The cerebral meninges has three layers:
 - i. **Dura** mater is the **superficial** layer of meninges
 - Tightly adherent to skull
 - Contains folds and **sinuses**
 - Contacts **endosteum** which lines the bones of the skull
 - ii. **Arachnoid** mater forms the middle of the three layers of meninges
 - iii. **Pia** mater is the delicate vascular fibrous membrane which is adherent to the brain.
 - B. **Brain** C710-C719
 - C. **Cranial nerves** C722-C729. See **Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves**
 - D. **Intracranial glands** **C751-C753**
 - i. Craniopharyngeal duct C752
 - ii. Pineal gland C753
 - iii. Pituitary gland C751

Continued on next page

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2. **Spinal** sites: includes the **spinal meninges** and **all** structures **within** the meninges (Intradural, within/in the spinal meninges).
 - A. **Spinal cord C720**
 - B. **Spinal meninges C701** covers/encloses the spinal nerve roots and the spinal cord.
 - C. Spinal nerve roots:
 - i. **Cervical** nerve, 8 pair, occipital nerve. Code to peripheral nerve of head, face, and neck C470
 - ii. **Coccygeal** nerve, 1 pair. Code to cauda equina C721
 - iii. **Lumbar** nerve, 5 pair. Code to cauda equina C721
 - iv. **Sacral** nerve, 5 pair. Code to cauda equina C721
 - v. **Thoracic** nerve, 12 pair. Code to peripheral nerves of thorax C473
3. Peripheral nerves
 - i. **Cervical** nerve, 8 pair, occipital nerve. Code to peripheral nerve of head, face, and neck C470
 - ii. **Thoracic** nerve, 12 pair. Code to peripheral nerves of thorax C473
 - iii. **Lumbar** nerve, 5 pair. Code to cauda equina C721
 - iv. **Sacral** nerve, 5 pair. Code to cauda equina C721
 - v. **Coccygeal** nerve, 1 pair. Code to cauda equina C721

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Table 2: Reportable Primary Sites

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

Site Group	Reportable Subsite Terms and Code
Brain	Brain NOS C719 Brain stem C717 Cerebellum NOS C716 Cerebrum C710 Frontal lobe C711 Occipital lobe C714 Overlapping lesion of brain C718 Parietal lobe C713 Temporal lobe C712 Ventricle NOS C715
Cranial Nerves	Abducent (cranial nerve VI) C725 Accessory (cranial nerve XI) C725 Acoustic (cranial nerve VIII) C724 Cranial nerve NOS C725 Facial (cranial nerve VII) 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 Trochlear (cranial nerve IV) C725 Vagus (cranial nerve X) C725

**Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

Site Group	Reportable Subsite Terms and Code
Ill-Defined Sites Central Nervous System	Nervous system NOS C729 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

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

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

Table 3 lists the more common histologies for CNS and Peripheral Nerves. For hematopoietic neoplasms such as lymphomas, myelomas, etc., see the [Hematopoietic Database](#).

Note: Behavior codes are not listed because all histologies are **malignant /3**.

Column 1 contains specific and NOS histology terms and codes.

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

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

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

Note: All tumors are malignant/invasive /3.

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term that encompasses a number of soft tissue tumors, including chondrosarcoma 9220/3 (column 3). Chondrosarcoma is also a NOS because it has a subtype/variant, mesenchymal chondrosarcoma 9240. The subtype/variant is indented under the NOS (chondrosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that chondrosarcoma has a subtype/variant.

When 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)

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Anaplastic ganglioglioma 9505		
Astroblastoma 9430		
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 ganglioneuroblastoma 9490		CNS embryonal tumor 9473
CNS neuroblastoma 9500		
Diffuse midline glioma H3 K27M mutant 9385*		
Embryonal carcinoma 9070		Yolk sac tumor 9071
Embryonal tumor with multilayered rosettes C19MC-altered 9478*	Embryonal tumor with multilayered rosettes, NOS ETMR	
Ependymoma 9391	Clear cell ependymoma Tancytic ependymoma	Anaplastic ependymoma 9392 Ependymoma, RELA fusion-positive 9396* Papillary ependymoma 9393
Epithelioid hemangioendothelioma 9133		
Germinoma 9064		

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
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
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Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Oligodendroglioma NOS 9450 <i>Note:</i> Oligodendroglioma NOS is used when molecular markers cannot fully be determined	Oligodendroglioma 1p/19q-codeleted Oligodendroglioma IDH-mutant Oligodendroglioma IDH-mutant and 1p/19q-codeleted	Anaplastic oligodendroglioma NOS 9451 IDH-mutant 1p/19q-codeleted IDH-mutant and 1p/19q-codeleted
Peripheral primitive neuroectodermal tumor 9364	Ewing sarcoma PpNET	
Pilocytic astrocytoma 9421 <i>Note:</i> ICD-O-3 lists a behavior code of /1. It is collected as /3 in North America.		Pilomyxoid astrocytoma 9425
Pineal parenchymal tumor of intermediate differentiation 9362	Pineoblastoma	Papillary tumor of the pineal region 9395
Sarcoma NOS 8800 <i>Note 1:</i> Chondrosarcoma 9220 has the following subtype/variant: Mesenchymal chondrosarcoma 9240 <i>Note 2:</i> Leiomyosarcoma 8890 has the following subtypes/variants: Epithelioid leiomyosarcoma 8891 Myxoid leiomyosarcoma 8896		Angiosarcoma 9120 Chondrosarcoma 9220 Mesenchymal chondrosarcoma 9240 Leiomyosarcoma/granular cell leiomyosarcoma/inflammatory leiomyosarcoma 8890 Epithelioid leiomyosarcoma 8891 Myxoid leiomyosarcoma 8896 Osteosarcoma 9180 Undifferentiated pleomorphic sarcoma/malignant fibrous histiocytoma 8802
Solitary fibrous tumor grade 3 8815	Hemangiopericytoma grade 3 Solitary fibrous tumor/Hemangiopericytoma grade 3 (CNS)	

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

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Table 4: Coding Primary Site for Malignant Tumors of Cranial and Peripheral Nerves

- Note 1:** Neoplasms arising in a **cranial** or **spinal** nerve are coded to the specific nerve in which it arises.
- Note 2:** Neoplasms, commonly meningiomas, arising in the **dura/meninges** of an **intracranial** nerve (cranial nerve within the skull) are coded to **cerebral meninges** C700.
- Note 3:** Neoplasms, commonly meningiomas, arising in the **dura/meninges** of the **spinal nerve roots** are coded to the ICD-O site code **spinal meninges** C701.
- Note 4:** It is important to **check the operative report** to determine whether the surgery is **intracranial** or **intradural**.

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

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

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

Column 3: Portions of the nerve coded to cranial nerve

Column 4: Portions of nerve coded to peripheral nerve

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

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Trochlear CN 4	Superior orbital fissure	Arises from the dorsal brain stem , loops around the brainstem and passes anteriorly within the subarachnoid space . It travels between the superior cerebellar and posterior cerebral arteries and through the dura , enters cavernous sinus C725	Enters the orbital fissure C470
Trigeminal CN 5 <i>Note:</i> Trigeminal is derived from Latin trigeminus which means born in threes (tri) and born at the same time (germinal). As the name implies, the nerve separates into three branches ; ophthalmic, maxillary, and mandibular	The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina. Superior orbital fissure. The foramen rotundum The foramen ovale.	CN5 originates in the pons . Upon leaving the pons it enters a small fossa posterior and inferolateral to the cavernous sinus called Meckel's (trigeminal) cave C725 .	<ul style="list-style-type: none"> • Ophthalmic nerve branch crosses the pterygopalatine fossa, inclines laterally on the back of the maxilla, and enters the orbit through the inferior orbital fissure where it is called the infraorbital nerve. It ends beneath the quadatus labii superius, and divides into multiple branches that spread to the side of the nose, the lower eyelid, and the upper lip C470 • Maxillary nerve leaves foramen rotundum and traverses the infraorbital groove and canal in the floor of the orbit, and appears on the face at the infraorbital foramen C470 • Mandibular nerve leaves via the foremen ovale travels along the mandibular groove C470

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Abducent CN 6	Cranial meninges	Exits brainstem at junction of pons and the medulla , enters the subarachnoid space and runs upward between the pons and the clivus entering the cavernous sinus C725	Dorello's canal and travels to the tip of the temporal bone . Enters orbit C470
Facial CN 7	Internal acoustic meatus	CN7 originates in the pons , along the posterior cranial fossa (posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity.) C725	Enters the temple through the internal auditory meatus and runs through the facial canal. C470
Acoustic or vestibulocochlear CN 8	Internal acoustic meatus	Originates in the brain stem (medulla oblongata) between the base of the brain (pons) and the spinal cord C724 Both the vestibular branch and the cochlear branch are located in the inner ear	
Glossopharyngeal CN 9	Jugular foramin ^a	Originates in the anterior portion of the medulla oblongata C725	Jugular foramen Between the 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

Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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Name and CN #	Exits Cranium Through	Site Code: Cranial Nerve	Site Code: Peripheral Nerve
Vagus CN 10	Jugular foramen	The vagus nerve originates from the medulla of the brainstem . C725	<p>CN10 descends within the carotid sheath medial to the internal jugular vein at the root of the neck C470.</p> <p>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</p> <p>Forms the anterior and posterior gastric nerves C475</p>
Accessory CN 11	Jugular foramen	The spinal accessory nerve originates in the neurons of the upper spinal cord, specifically C1-C5/C6 spinal nerve roots . The nerve enters the foramen magnum or lateral aspect of the medulla oblongata . The fibers of the spinal accessory nerve coalesce to form spinal rootlets, roots , and finally the spinal accessory nerve itself C725	<p>The nerve exits the skull through the jugular foramen. It then runs along the internal carotid artery within the neck C470</p> <p>Reaches the sternocleidomastoid muscle and the trapezius C476</p>
Hypoglossal CN 12	Hypoglossal canal	CN12 starts in the hypoglossal nucleus of the brainstem , C725	CN12 exits the hypoglossal canal, traveling between the carotid artery and jugular vein, ending under the tongue C470

**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)**

Section 3: Additional Information to Complete the Abstract

Conflicting Information on Pathology Report(s)

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

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

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

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

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

**Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

Table 5: Paired Sites

Use **Table 5** to identify sites for which laterality **must** be coded. Do **not** use this table to determine multiple primaries.

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

Note 1: Midline tumors are common for glioblastoma multiform and meningiomas.

Note 2: SEER allows laterality to be coded for sites other than those in the table.

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

Table 6: Non-malignant CNS Tumors with the Potential of Transforming to Malignant Behavior

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

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

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

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

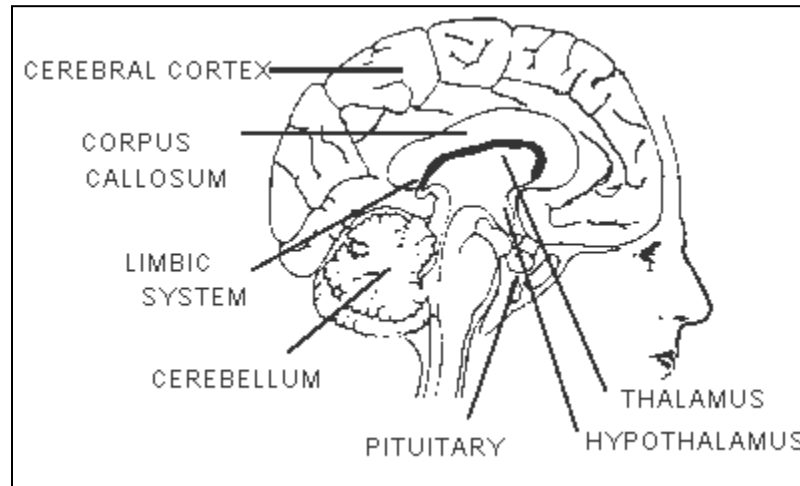
Column 1 is the **non-malignant** ICD-O histology term and code.

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

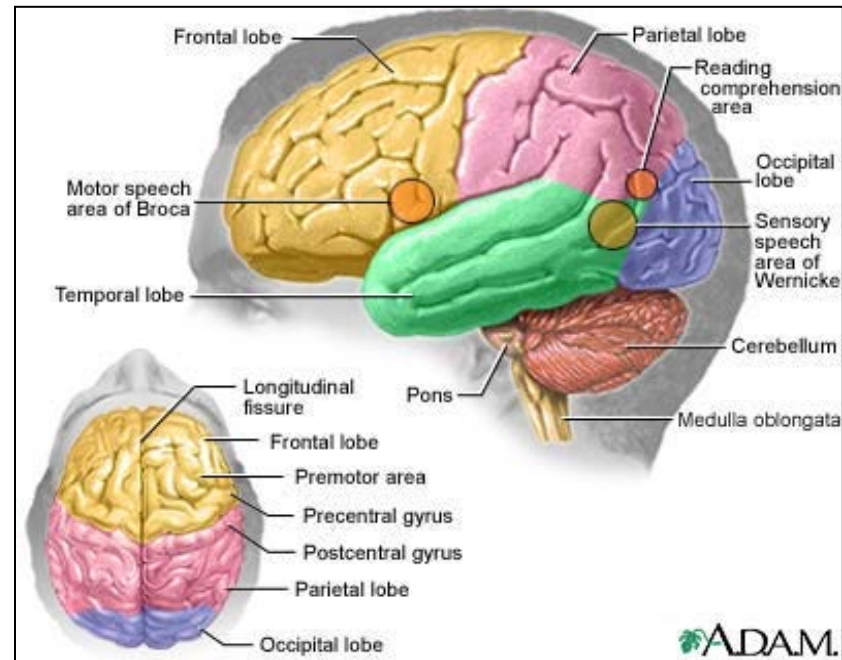
Original Histology and Code	Transformed Histology and Code
Chondroma 9220/0	Chondrosarcoma 9220/3
Ganglioglioma 9505/1	Anaplastic ganglioglioma 9505/3
Hemangioma 9120/0	Angiosarcoma 9120/3
Hemangiopericytoma 9150/1	Anaplastic hemangiopericytoma 9150/3
Leiomyoma 8890/0	Leiomyosarcoma 8890/3
Lipoma 8850/0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

**Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)**

Illustrations

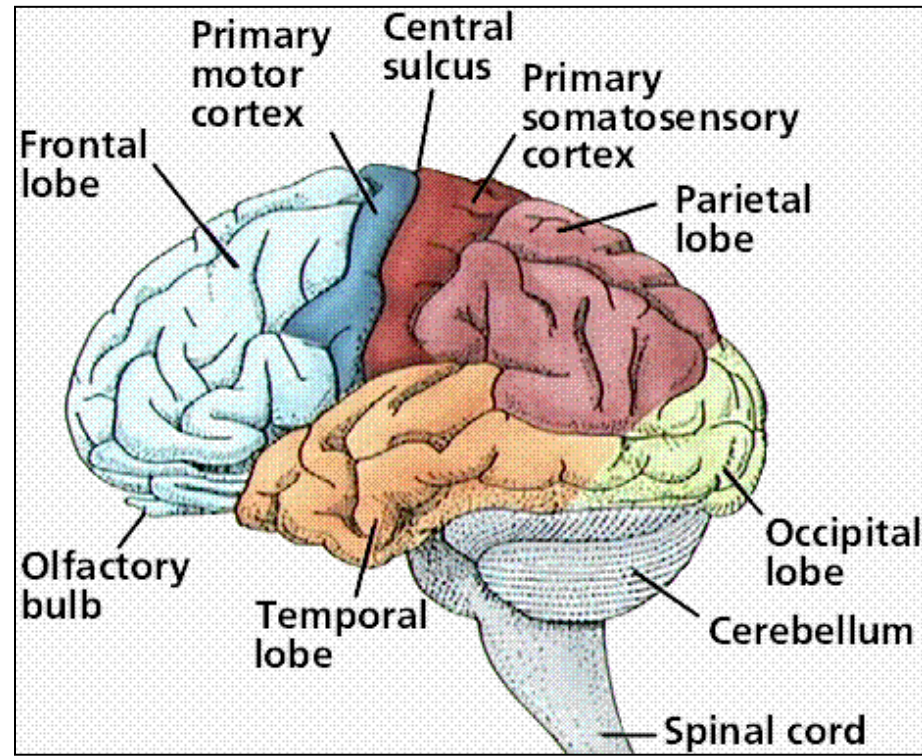


Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
C470-C479, C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
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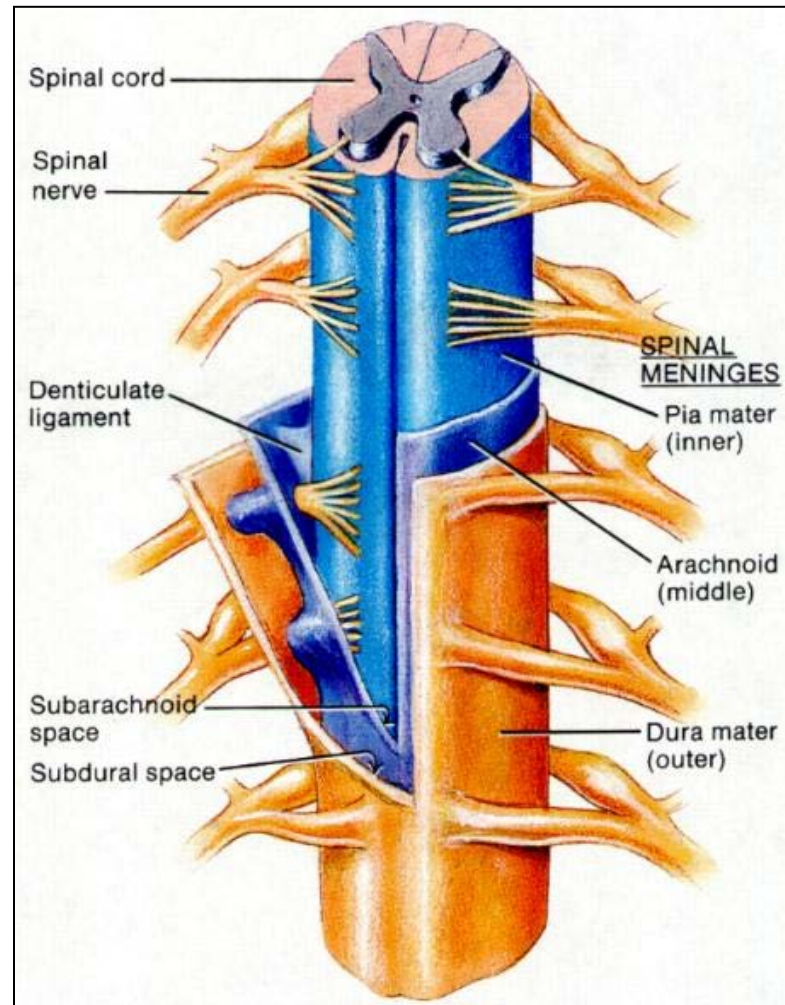
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Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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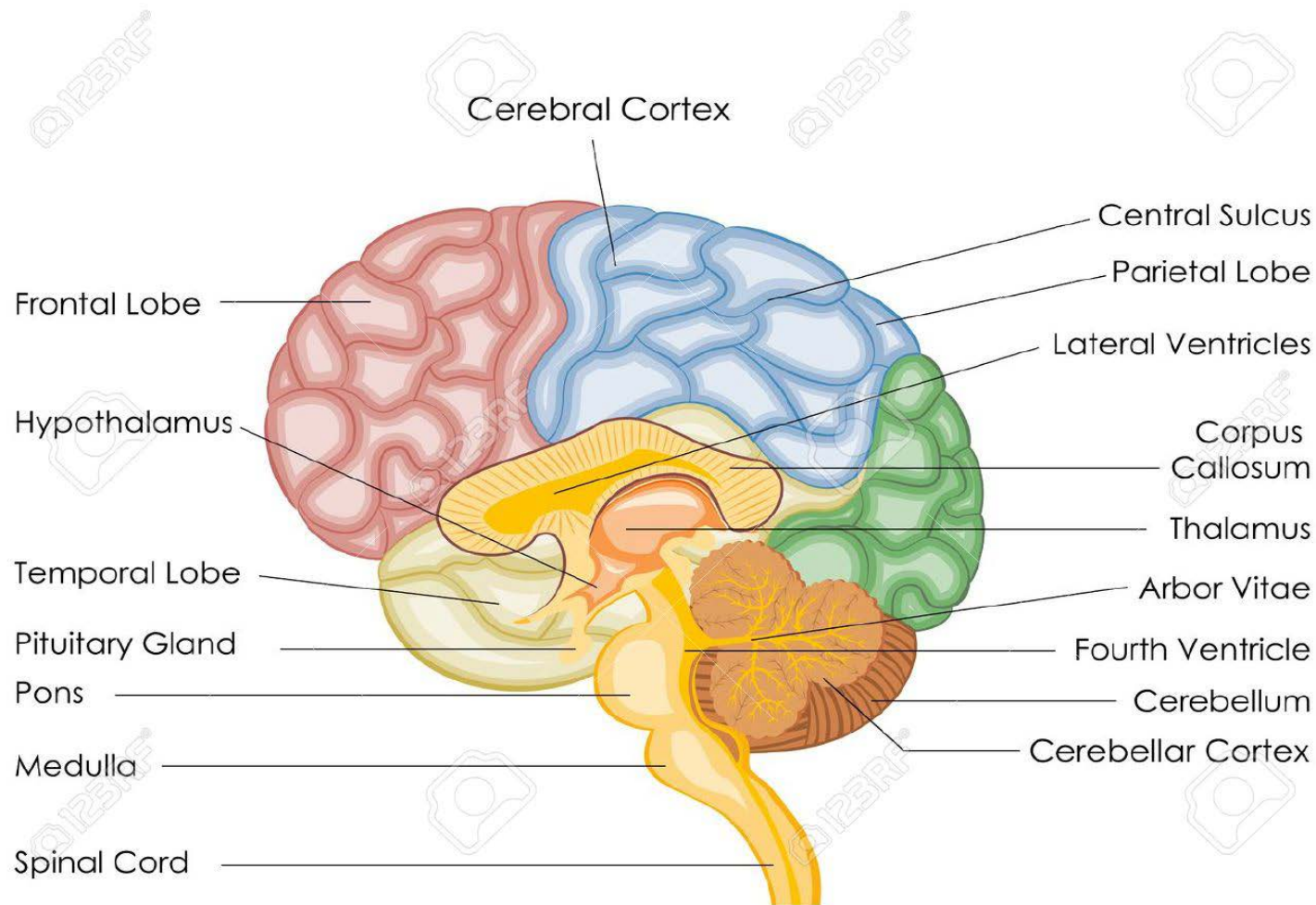


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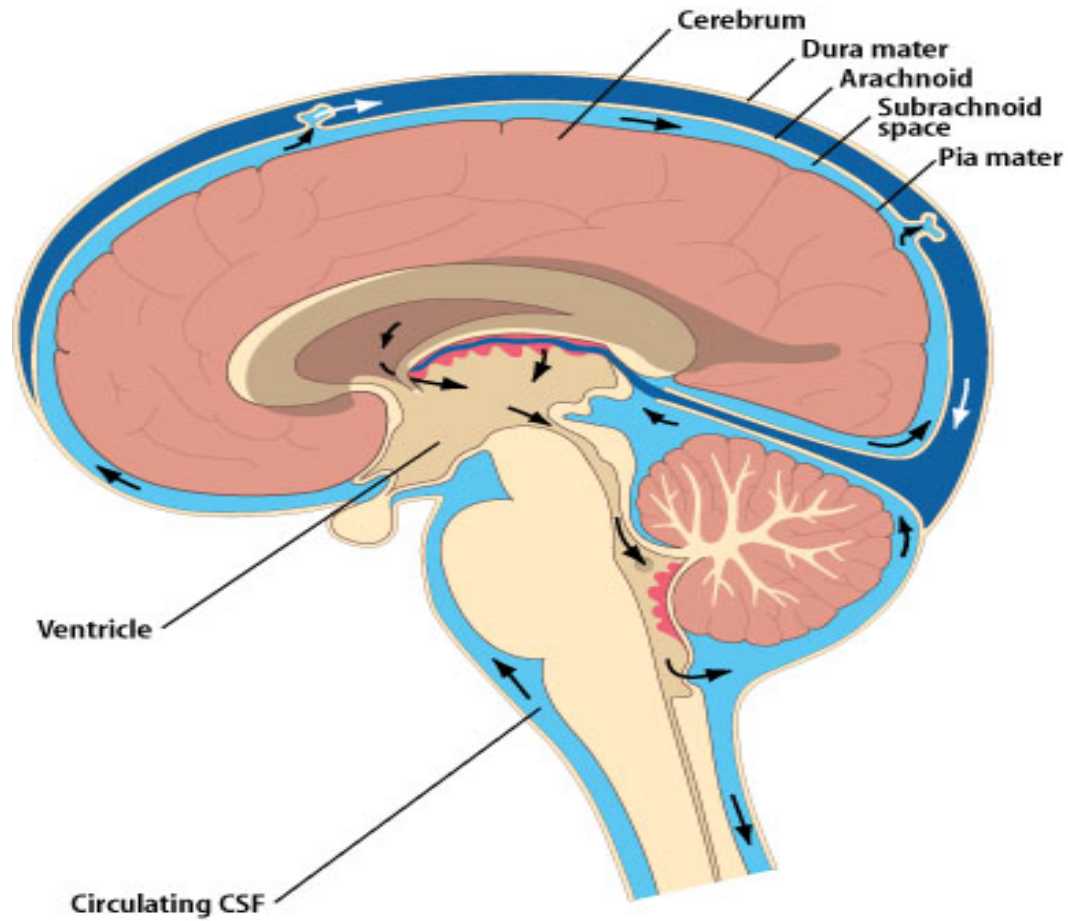
Malignant CNS and Peripheral Nerves Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



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**ⁱ when it is not possible to determine if there is a **single tumor or multiple tumors**.

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

Note 2: Examples of cases with minimal information include

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

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

ⁱPrepare one abstract. Use the [histology rules](#) to assign the appropriate histology code.

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

Single Tumor

IMPORTANT: The **major difference** between **M4** and **M5** is:
M4: No resection as first course of treatment **AND** when the same tumor is subsequently **resected, pathology proves malignant** behavior
M5: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo**) is **malignant**

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

Note 1: A single tumor is always a single primary.

Note 2: The tumor **may overlap onto** or **extend** into **adjacent**/contiguous site or subsites.

Note 3: The tumor may have two or more histologic components.

Rule M3 Code a **single primary**ⁱ when a neoplasm is **originally diagnosed** as an **oligodendroglioma** and subsequently **recurs** in residual tumor tissue with **different features** such as a densely cellular tumor with pseudo palisading necrosis.

Note 1: The pathology may state that the recurrence “looks like” or “has the appearance of” a glioblastoma multiforme (GBM). This is not a true GBM.

Note 2: Record as a recurrence for those registrars who collect recurrence data.

Rule M4 Abstract a **single primary**ⁱ (the malignant) when a single tumor meets the following two criteria:

1. The original diagnosis was non-malignant /0 or /1 **AND**
 - o 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.

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

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

Multiple Tumors

Note 1: Multiple tumors may be a single primary or multiple primaries.

Note 2: Separate, non-contiguous tumors are always multiple primaries when:

- In the CNS (see [Table 2](#)) AND in a site other than the CNS

Example: Patient has a malignant nerve sheath tumor of the right arm C471 and non-metastatic adenocarcinoma of the lung. The lung is not a CNS site. Abstract two primaries.

- In different CNS sites (see Rule M8)

IMPORTANT: The major difference between **M4** and **M5** is:

M4: No resection as first course of treatment **AND** when the same tumor is subsequently **resected**, **pathology proves malignant** behavior

M5: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo**) is **malignant**

Rule M5 Abstract **multiple primaries**ⁱⁱ when there are multiple CNS tumors, one of which is malignant /3 and the other is non-malignant /0 or /1.

- Original non-malignant tumor followed by malignant tumor
 - o Patient had a resection of the non-malignant tumor (not the same tumor) **OR**
 - o It is unknown/not documented if the patient had a resection
- Simultaneous non-malignant and malignant tumors
 - o Abstract both the malignant and the non-malignant tumors

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

Note 2: See [Table 2](#) in the Equivalent Terms and Definitions for a listing of CNS sites.

Note 3: A non-malignant CNS tumor and a malignant CNS tumor are **always multiple** primaries (timing and primary sites are irrelevant). Prepare two abstracts; one for the non-malignant and another for the malignant tumor.

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

Rule M6 Abstract **multiple primaries**ⁱⁱ when a patient has a glial tumor and is subsequently diagnosed with a **glioblastoma multiforme 9440** (GBM).

Note 1: Definition of a glial tumor: Any tumor arising from the CNS glial cells. The following is simply a list of all tumors which would be classified as glial.

- Astroblastoma **9430**
- Astrocytomas **9400** and all subtypes
 - Anaplastic astrocytoma IDH-mutant/wildtype; anaplastic astrocytoma NOS **9401**
 - Gemistocytic astrocytoma IDH-mutant **9411**
- Diffuse midline glioma H3 K27M Mutant **9385**
- Ependymoma **9391** and all subtypes
 - Anaplastic ependymoma **9392**
 - Ependymoma, RELA fusion-positive **9396**
 - Papillary ependymoma **9393**
- Glioblastoma **9440** and all subtypes (**this is a glial tumor; however do not apply this rule to a GBM followed by a GBM**)
 - Giant cell glioblastoma **9441**
 - Glioblastoma IDH-mutant **9445**
 - Gliosarcoma **9442**
- Oligodendroglioma and all subtypes **9450**
 - Anaplastic oligodendroglioma; IDH-mutant; 1p/19q-codeleted; IDH-mutant and 1p/19q-codeleted **9451**
- Pleomorphic xanthroastrocytoma **9424**

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

Note 3: Abstracting GBM as a new primary will allow analysis of:

- The number of tumors that recur as a more aggressive histology (GBM)
- The time interval between occurrence of the glial or astrocytic tumors and a GBM
- Which histologies are more likely to recur as a GBM

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

Rule M7 Abstract a **single primary**ⁱ when there are separate, non-contiguous tumors in the brain (multicentric/multifocal) with the **same histology XXXX**. Tumors may be any of the following combinations:

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

Example: The patient had a resection of an anaplastic astrocytoma 9401 in the right parietal lobe. Three months later the patient is diagnosed with a de novo anaplastic astrocytoma in the left parietal lobe. This is one primary because neither laterality nor timing are used to determine multiple primary status.

Note 1: **Multiple sites/subsites** and/or different **lateralities** **imply** either **metastatic** or **multifocal/multicentric** disease. Glioblastoma multiforme commonly exhibits multiple tumors which are described as **multifocal/multicentric**.

Note 2: Metastases are never used to determine multiple primaries. **Seeding metastasis** is often noted for the following tumors:

- Glioblastoma multiforme
- pNET-medulloblastoma
- Oligodendroglioma

Note 3: **Hereditary** syndromes frequently exhibit multiple tumors including the following:

- Neurofibromatosis type 1 (NF1)
 - Malignant peripheral nerve sheath tumors (MPNST)
- Neurofibromatosis type 2 (NF2)
 - Anaplastic ependymomas
 - Meningiomas

Note 4: Most malignant neoplasms are **single tumors** with the exception of those listed in this rule.

Note 5: This is a **change** from/**clarification** to previous rules.

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

Rule M8 Abstract **multiple primaries**ⁱⁱ when multiple tumors are present in any of the following **sites** or **subsites**:

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

Rule M9 Abstract **multiple primaries**ⁱⁱ when separate, non-contiguous tumors are two or more **different subtypes/variants** in Column 3, **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS:** Anaplastic astrocytoma IDH-mutant 9401 and gemistocytic astrocytoma IDH-mutant 9411 are both subtypes of astrocytoma NOS 9400/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS:** Papillary ependymoma 9393 is a subtype of ependymoma NOS 9391; gliosarcoma 9442 is a subtype of glioblastoma NOS 9440. They are distinctly different histologies. Abstract multiple primaries.

Rule M10 Abstract a **single primary**ⁱ when separate, non-contiguous tumors are on **the same row** in **Table 3** in the Equivalent Terms and Definitions. Timing is irrelevant.

Note: The same row means the tumors are:

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

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.

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

Note: Non-malignant CNS tumors have a separate set of rules.

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed prior to neoadjuvant treatment.
Note 1: Histology changes 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/documented

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

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)

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

Example 1: The pathology diagnosis is astrocytoma consistent with gemistocytic astrocytoma IDH-mutant. The oncology consult says the patient has gemistocytic astrocytoma IDH-mutant in the occipital lobe. This is clinical confirmation of the diagnosis, code gemistocytic astrocytoma IDH-mutant. The case meets the criteria in bullet 1.

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

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

List of Ambiguous Terminology

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

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

- Architecture
- Foci; focus; focal
- Pattern

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

Single Tumor

- Rule H1** Code the **reportable CNS tumor** ([Table 3](#) in the Equivalent Terms and Definitions) when a patient has any of the following:
- Neurofibromatosis type 1 (NF1)
 - Neurofibromatosis type 2 (NF2)
 - **Schwannomatosis**
- Note 1:* Do not code NF1 or NF2 as neurofibromatosis. NF1, NF2, and schwannomatosis are genetic syndromes which have a high risk of developing reportable and non-reportable tumors. **ONLY** abstract **reportable tumors** such as malignant peripheral nerve sheath tumors.
- Note 2:* Tumors are reportable when they meet the behavior (/3) and histology requirements (see [Reportability Criteria](#)).
- Note 3:* Schwannomatosis is a newer term for a distinct subtype/variant of the genetic diseases NF1 and NF2.
- Example:* Patient presents with vestibular schwannoma (acoustic neuroma). Genetic testing proves the patient has NF2. Report the acoustic nerve neuroma.
- Rule H2** Code malignant meningioma **9530** when the diagnosis specifically states malignant/invasive.
- Note:* Code the behavior designated in the pathology report. Do not code malignant /3 based on invasion of brain or skull. Atypical meningioma (non-malignant) may invade contiguous structures.
- Rule H3** Code the histology when only **one histology** is present.
- Note 1:* Use [Table 3](#) to code histology. New codes, terms, and synonyms are included in [Table 3](#) and coding errors may occur if the table is not used.
- Note 2:* When the histology is **not listed** in [Table 3](#), use the **ICD-O** and all **updates**.
- Note 3:* Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in [Table 3](#), ICD-O or all updates.

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 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
- Intraspidal; occurring within the spinal column especially the vertebral canal
- Majority; major; predominantly; greater than 50%
- Non-malignant is synonymous with:
 - /0 Benign
 - /1 Uncertain whether benign or malignant; borderline malignancy; low malignant potential; uncertain malignant potential
 - WHO Grade 1
- Site; topography
- Subarachnoid space; between the arachnoid mater and the pia mater of spinal meninges
- Type; subtype; variant
- Tumor; mass; tumor mass; lesion; neoplasm
 - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician’s statement** that the term is a non-malignant tumor/neoplasm
 - These terms are used **ONLY** for determining multiple primaries
 - **DO NOT USE** these terms for **casefinding** or **determining reportability**

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).

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)

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](#)).

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)

Directory of Sections and Tables

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 Groups**
- C. **Table 2:** Reportable Primary Sites
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- 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

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

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

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- C. PET
- D. Angiogram

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

Table 1: WHO Grades of Select CNS Neoplasms

Note 1: CNS WHO classifications use a grading scheme that is a "malignancy scale" ranging across a wide variety of neoplasms rather than a strict histologic grading system that can be applied equally to all tumor types.

Note 2: See the SEER and CoC Manuals for instructions on coding grade for CNS tumors.

Note 3: The table **does not** contain all neoplasms that may occur in the CNS.

WHO Grade Definitions

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

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

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
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Histology	WHO Grade
Anaplastic (malignant) meningioma	3
Anaplastic astrocytoma, IDH mutant	3
Anaplastic ependymoma	3
Anaplastic ganglioglioma	3
Anaplastic oligodendroglioma IDH mutant and 1p/19q-codeleted	3
Anaplastic pleomorphic xanthroastrocytoma	3
Angiocentric glioma	1
Atypical choroid plexus papilloma	2
Atypical meningioma	2
Atypical teratoid/rhabdoid tumor	4
Central neurocytoma	2
Cerebellar liponeurocytoma	2
Chordoid glioma of third ventricle	2
Choroid plexus carcinoma	3
Choroid plexus papilloma	1
CNS embryonal tumor NOS	4
CNS embryonal tumor with rhabdoid features	4
Craniopharyngioma	1
Desmoplastic infantile astrocytoma and ganglioglioma	1
Diffuse astrocytoma, IDH mutant	2
Diffuse midline glioma, H3K27M-mutant	4
Dysembryoplastic neuroepithelial tumor	1
Dysplastic gangliocytoma of cerebellum (D=Lhemitte-Duclos)	1
Ependymoma	2
Ependymoma, RELA fusion-positive	2 or 3

Non-Malignant CNS Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Histology	WHO Grade
<i>Note:</i> Tissue/pathology reports or CAP report will specify whether this is WHO Grade 2 or 3	
Extraventricular neurocytoma	2
Gangliocytoma	1
Glioblastoma, IDH mutant	4
Glioblastoma, IDH wildtype	4
Granular cell tumor	1
Hemangioblastoma	1
Malignant peripheral nerve sheath tumor (MPNST)	2, 3, or 4
<i>Note:</i> Tissue/pathology reports or CAP report will specify whether this is WHO Grade 2, 3, or 4	
Medulloblastoma (including all subtypes)	4
Medulloepithelioma	4
Meningioma	1
Myxopapillary ependymoma	1
Neurofibroma	1
Oligodendroglioma IDH mutant and 1p/19q-codeleted	2
Papillary glioneuronal tumor	1
Papillary tumor of the pineal region	2 or 3
<i>Note:</i> Tissue/pathology report or CAP will specify whether it is WHO Grade 2 or 3	
Perineuroma	1
Pilocytic astrocytoma	1
<i>Note:</i> ICD-O-3 lists a behavior code of /1. US and Canada collect as /3.	
Pineal parenchymal tumor of intermediate differentiation	2 or 3
<i>Note:</i> Tissue/pathology or CAP synoptic report will specify WHO Grade 2 or 3	

Non-Malignant CNS Equivalent Terms and Definitions
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Histology	WHO Grade
Pineoblastoma	4
Pineocytoma	1
Pituicytoma	1
Pleomorphic xanthroastrocytoma	2
Rosette-forming glioneuronal tumor	1
Schwannoma	1
Solitary fibrous tumor/hemangiopericytoma	1, 2, or 3
<i>Note:</i> Tissue/pathology will specify WHO Grade 1, 2, or 3	
Spindle cell oncocytoma	1
Subependymal giant cell astrocytoma	1
Subependymoma	1

Non-Malignant CNS Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Section 2: Reportable Primary Sites and Histologies

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

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

- **Intraosseous**

Note: The **dura** layer of the meninges **contacts** the **endosteum** of the bones of the skull. The primary site for intraosseous meningioma is cranial meninges C700.

- **Sphenoid wing**

Note 1: Sphenoid wing meningiomas arise in the **cranial meninges** C700 which **covers** the bony structure called the sphenoid wing.

Note 2: The term “sphenoid wing meningioma” is used to identify the **location** of the meningioma because sphenoid wing meningiomas may be very **invasive**, spreading to the dura of the frontal, temporal and orbital regions.

- **Cavernous sinus**

Note 1: Cavernous sinus is located between the endosteal and meningeal layers of the dura.

Note 2: There is **no ICD-O site** code for cavernous sinus because tumors do not arise in the sinus itself; the primary sites are:

- The **cranial nerves** passing through the sinus (trochlear, abducent **C725**) **OR**
- The cerebral **meninges/dura C700** covering the cranial nerve

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

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

Non-Malignant CNS Equivalent Terms and Definitions
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(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

Reportable Primary Site Groups

The two major **groups** of reportable sites are:

1. **Intracranial** (within the skull/cranium)
2. **Spinal sites** (spinal meninges and sites within the spinal meninges)

Reportable Primary Sites and their ICD-O Codes

1. Intracranial (intra means inside or within; intracranial is inside the skull).
 - A. **Cerebral/cranial dura/meninges C700**. The cerebral meninges has three layers:
 - i. **Dura** mater is the **superficial** layer of meninges
 - Tightly adherent to skull
 - Contains folds and **sinuses**
 - Contacts **endosteum** which lines the bones of the skull
 - ii. **Arachnoid** mater forms the middle of the three layers of meninges
 - iii. **Pia** mater is the delicate vascular fibrous membrane which is adherent to the brain.
 - B. **Brain C710-C719**
 - C. **Cranial nerves C722-C729**. See [Table 3: Reportability of Non-Malignant Cranial Nerve \(CN\) Tumors](#)
 - D. **Intracranial glands C751-C753**
 - i. Craniopharyngeal duct C752
 - ii. Pineal gland C753
 - iii. Pituitary gland C751
2. **Spinal sites**: includes the **spinal meninges** and **all** structures **within** the meninges (Intradural, within/in the spinal meninges).
 - A. Spinal cord **C720**
 - B. The spinal meninges **C701** covers/encloses the spinal cord.

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 2: Reportable Primary Sites

Column 1 lists the reportable primary site term.

Column 2 lists the site code for the reportable primary site.

Site Group	Reportable Subsite Terms and Code
Brain	Brain NOS C719 Brain stem C717 Cerebellum NOS C716 Cerebrum C710 Frontal lobe C711 Occipital lobe C714 Overlapping lesion of brain and central nervous system C718 Parietal lobe C713 Temporal lobe C712 Ventricle NOS C715
Cranial Nerves	Abducent (cranial nerve VI) C725 Accessory (cranial nerve XI) C725 Acoustic (cranial nerve VIII) C724 Cranial nerve NOS C725 Facial (cranial nerve VII) 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 Trochlear (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)

Site Group	Reportable Subsite Terms and Code
Ill-Defined Sites Central Nervous System	Nervous system NOS C729 Overlapping lesion of brain and central nervous system C728
Intracranial Duct and Glands	Craniopharyngeal duct C752 Pineal gland C753 Pituitary gland C751
Meninges	Cerebral meninges C700 Meninges NOS C709 Spinal meninges C701
Spinal Sites	Cauda equina/conus medullaris/filum terminale C721 Meninges NOS C709 Spinal cord C720 Spinal meninges C701

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 3: Reportability of Non-Malignant Cranial Nerve (CN) Tumors

This table is used to determine whether non-malignant cranial nerve tumors and cranial nerve meningiomas are **reportable**. When cranial nerves **exit** the **intracranial** space, they become **peripheral nerves (non-reportable)**.

Note 1: A neoplasm arising in a **cranial** nerve is coded to the **specific nerve** in which it **arises**.

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

Note 3: It is important to check the **operative report** to determine whether the surgery is **intracranial** or **intradural**.

Note 4: This table is used for **non-malignant** neoplasms **ONLY**.

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

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

Column 3: **Reportable** portions of the cranial nerve. Of note, malignant neoplasms are reportable for all parts of the cranial nerves

Column 4: **Non-reportable** portions of the cranial nerve

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

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

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Trochlear CN 4 C725	Superior orbital fissure	Arises from the dorsal brain stem , loops around the brainstem and passes anteriorly within the subarachnoid space . It travels between the superior cerebellar and posterior cerebral arteries and through the dura , enters cavernous sinus .	Enters the orbital fissure .
Trigeminal CN 5 C725 <i>Note:</i> Trigeminal is derived from Latin trigeminus which means born in threes (tri) and born at the same time (germinal). As the name implies, the nerve separates into three branches ; ophthalmic, maxillary, and mandibular	The ophthalmic, maxillary and mandibular branches leave the skull through three separate foramina, the superior orbital fissure, the foramen rotundum and the foramen ovale.	CN5 originates in the pons . Upon leaving the pons it enters a small fossa posterior and inferolateral to the cavernous sinus called Meckel's (trigeminal) cave .	<ul style="list-style-type: none"> • Ophthalmic nerve branch crosses the pterygopalatine fossa, inclines laterally on the back of the maxilla, and enters the orbit through the inferior orbital fissure where it is called the infraorbital nerve. It ends beneath the quadatus labii superius, and divides into multiple branches that spread to the side of the nose, the lower eyelid, and the upper lip • Maxillary nerve leaves foramen rotundum and traverses the infraorbital groove and canal in the floor of the orbit, and appears on the face at the infraorbital foramen. • Mandibular nerve leaves via the foremen ovale travels along the mandibular groove

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

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Abducent CN 6 C725	Cranial meninges	Exits brainstem at junction of pons and the medulla , enters the subarachnoid space and runs upward between the pons and the clivus entering the cavernous sinus .	Dorello's canal and travels to the tip of the temporal bone . Enters orbit
Facial CN 7 C725	Internal acoustic meatus	CN7 originates in the pons , along the posterior cranial fossa (the posterior cranial fossa is part of the intracranial cavity)	Enters the temple through the internal auditory meatus and runs through the facial canal .
Acoustic or vestibulocochlear CN 8 C724	Internal acoustic meatus	Originates in the brain stem (medulla oblongata) between the base of the brain (pons) and the spinal cord Both the vestibular branch and the cochlear branch are located in the inner ear	
Glossopharyngeal CN 9 C725	Jugular foramen	Originates in the anterior portion of the medulla oblongata	Jugular foramen Between the 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

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

Name and CN #	Exits Cranium Through	Reportable Portions of CN	Non-Reportable Portions of CN
Vagus CN 10 C725	Jugular foramen	The vagus nerve originates from the medulla of the brainstem .	CN10 descends within the carotid sheath medial to the internal jugular vein at the root of the neck . The right vagus crosses in front of the subclavian artery and travels into the fat behind the blood vessels , reaching the thorax . It then inclines behind the hilum of the right lung and moves toward the esophagus. The nerve splits into the right and left vagus at the esophageal plexus forming the anterior and posterior gastric nerves
Accessory CN 11 C725	Jugular foramen	The nerve enters the foramen magnum or lateral aspect of the medulla oblongata .	<p>The spinal accessory nerve originates in the neurons of the upper spinal cord, specifically C1-C5/C6 spinal nerve roots.</p> <p>The fibers of the spinal accessory nerve coalesce to form spinal rootlets, roots, and finally the spinal accessory nerve itself.</p> <p>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.</p>

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

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

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

Table 4: Non-Reportable Neoplasms

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

Non-reportable Histology Term	Non-reportable Histology Code	Definitions and Sites
Carcinomas	8010-8060, 8071-8671, 8940-8941	Brain C710-C719 Site/histology edit carcinomas/brain
Carcinomas	8010-8671, 8940-8941	Cerebral meninges, spinal meninges, meninges NOS C700-C709 Site/histology edit carcinomas/meninges
Carcinomas	8010-8671, 8940-8941	C721-C729 (Other central nervous system) Site/histology edit carcinomas/other CNS
Colloid cyst	No code	
Epidermoid tumor/cyst	No code	
Glomus tympanicum, glomus jugulare	8690/1	These tumors occur in the inner ear, the aortic body and other paraganglia respectively; these sites are not reportable .
Hygroma	9173/0	
Hypothalamic hamartoma	No code	Occurs in hypothalamus
Neurofibromatosis, 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

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

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Histology Term and Code	Most Common Intracranial Primary Site
Pilocytic astrocytoma/juvenile pilocytic astrocytoma 9421/1	Posterior fossa C719 , cerebrum C710
Pineocytoma 9361/1	Pineal gland C753
Pituicytoma 9432/1*	Pituitary gland C751 , sella turcica C751 , suprasellar C719
Pituitary adenoma 8272/0	Pituitary gland C751
Prolactinoma 8271/0	Pituitary gland C751
Subependymal giant cell tumor (SEGA) 9384/1	Lateral ventricles C715
Subependymoma 9383/1	Intraventricular site (lateral/third ventricle, rare C715 and IV ventricle C717)

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

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

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

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

Column 1 contains specific and NOS histology terms.

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

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

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

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Angiocentric glioma 9431/1*	Angiocentric neuroepithelial tumor Monomorphous angiocentric glioma	
Benign fibrous histiocytoma 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)

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
Desmoplastic infantile astrocytoma and ganglioglioma 9412/1		
Dysembryoplastic neuroepithelial tumor 9413/0	DNT	
Gangliocytoma 9492/0		Dysplastic cerebellar gangliocytoma/Lhermitte-Duclos disease 9493/0
Ganglioglioma 9505/1		
Granular cell tumor of the sellar region 9582/0		
Hemangioblastoma 9161/1	Capillary hemangioblastoma	
Hemangioma 9120/0		Cavernous hemangioma 9121/0
Leiomyoma 8890/0		
Lipoma 8850/0		Hibernoma 8880/0
Meningeal melanocytosis 8728/0		Meningeal melanocytoma 8728/1
Meningioma 9530/0	Lymphoplasmacyte-rich meningioma Metaplastic meningioma Microcystic meningioma Secretory meningioma	Angiomatous meningioma 9534/0 Atypical meningioma 9539/1 Clear cell/chordoid meningioma 9538/1 Fibrous meningioma 9532/0 Meningothelial meningioma 9531/0 Psammomatous meningioma 9533/0 Transitional meningioma 9537/0
Myofibroblastoma 8825/0		Inflammatory myofibroblastic tumor 8825/1
Myxopapillary ependymoma 9394/1		
Neurofibroma 9540/0	Atypical neurofibroma	Plexiform neurofibroma 9550/0
Optic glioma/pilocytic astrocytoma 9421/1		
Osteoma 9180/0		

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

NOS/Specific Histology Term and Code	Synonyms	Subtypes/Variants Histology Term and Codes
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		

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)

Section 3: Additional Information to Complete Abstract

Conflicting Information on Pathology Report(s)

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

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

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

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

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

Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – 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.

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

Note 1: Midline tumors are most common for the following histologies: glioblastoma multiforme, meningiomas, lymphomas (usually located near the midline), and epidermoid cysts (which can cross the midline via the subarachnoid space).

Note 2: SEER allows laterality to be coded for sites other than those in the table.

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

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

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

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

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

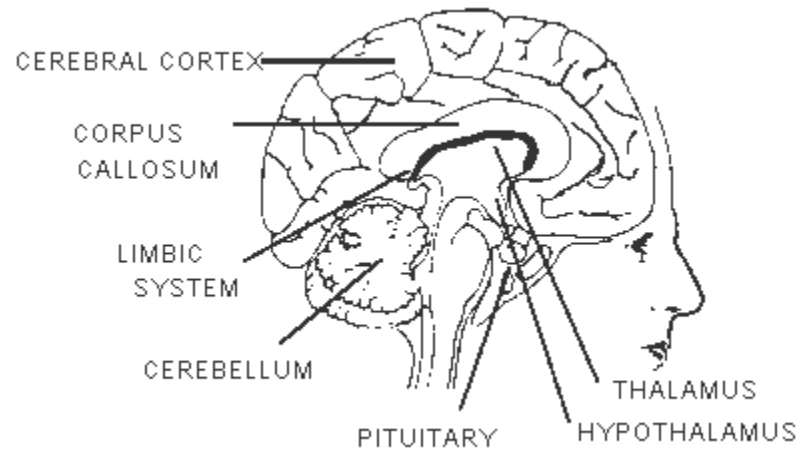
Column 1 is the **non-malignant** ICD-O histology term and code.

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

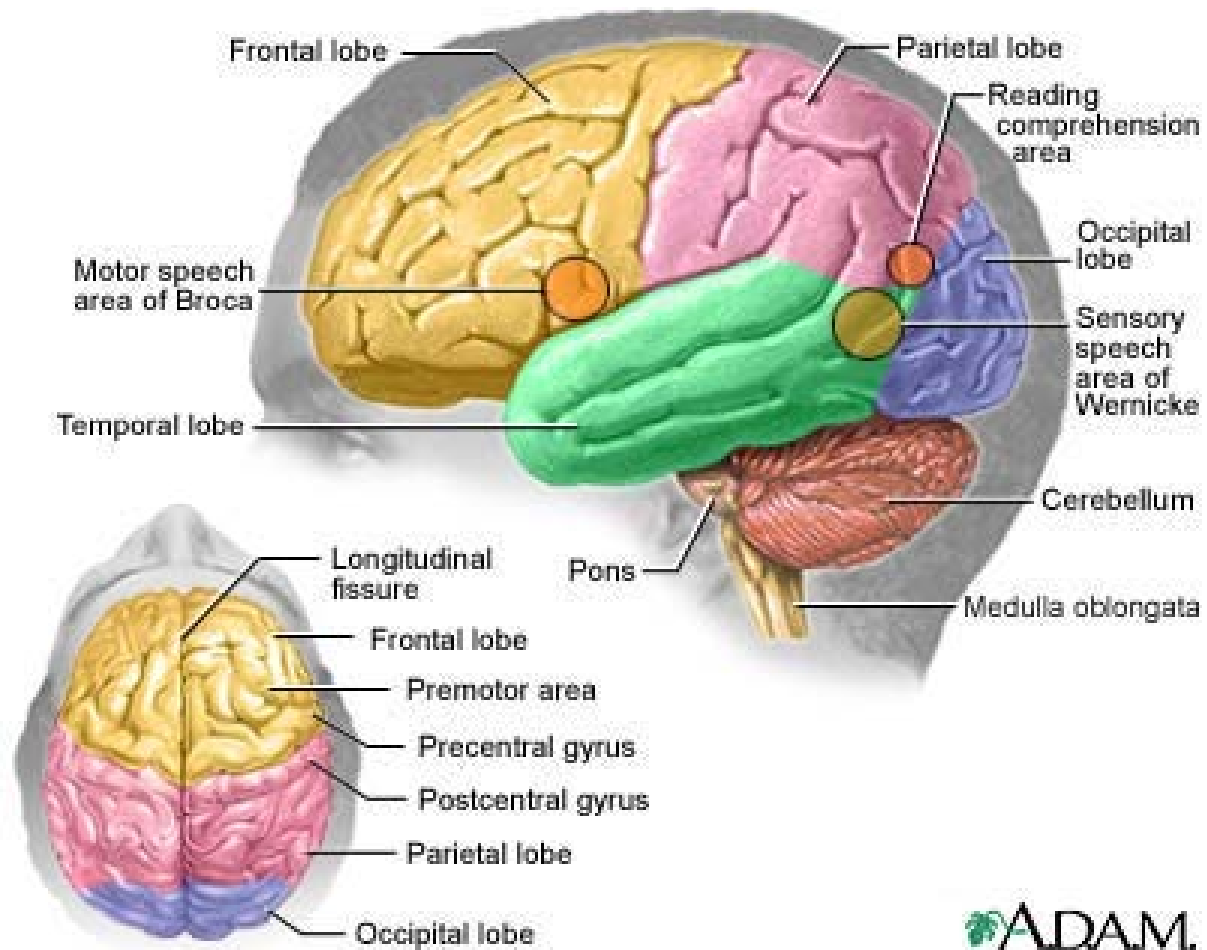
Original Histology and Code	Transformed Histology and Code
Chondroma 9220/0	Chondrosarcoma 9220/3
Ganglioglioma 9505/1	Anaplastic ganglioglioma 9505/3
Hemangioma 9120/0	Angiosarcoma 9120/3
Hemangiopericytoma 9150/1	Anaplastic hemangiopericytoma 9150/3
Leiomyoma 8890/0	Leiomyosarcoma 8890/3
Lipoma 8850/0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

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)

Illustrations



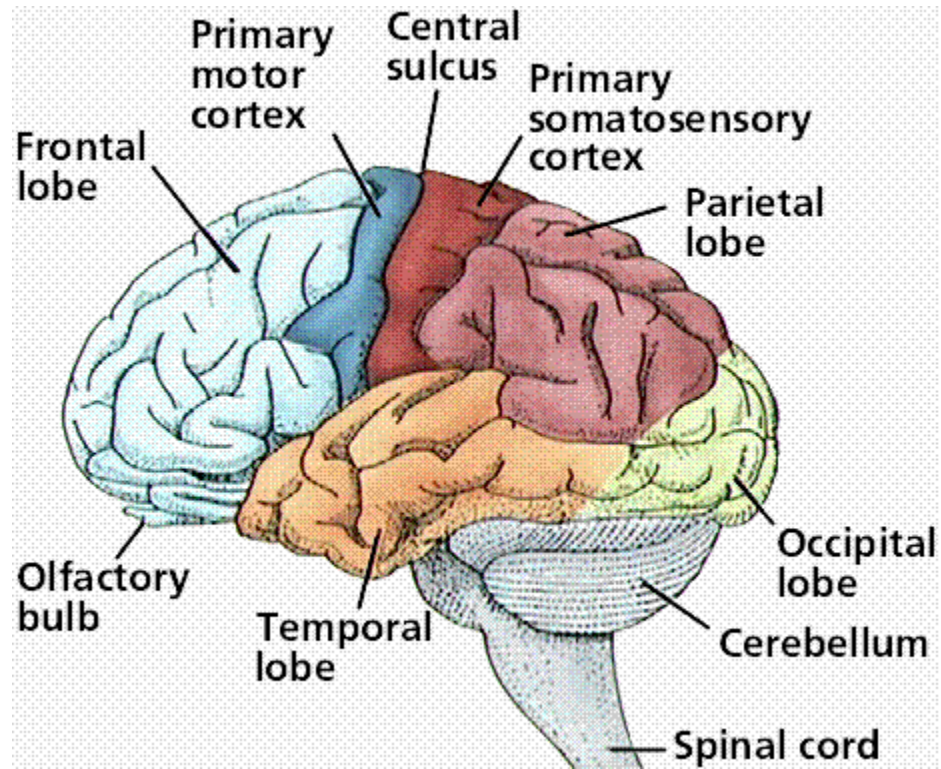
Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



 ADAM

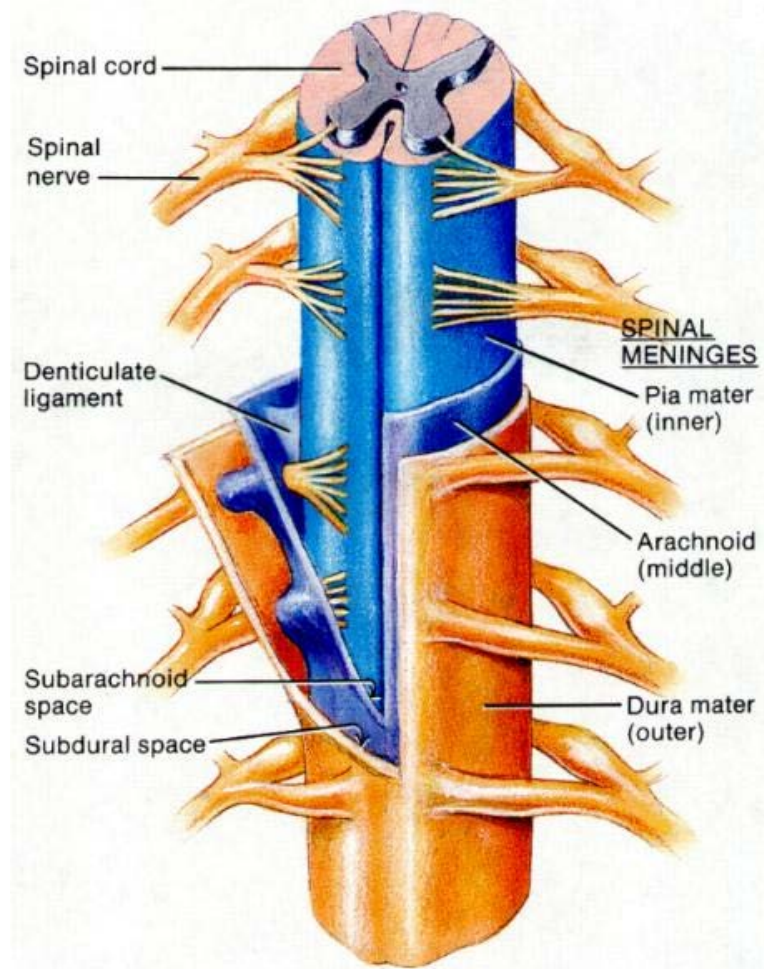
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Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)

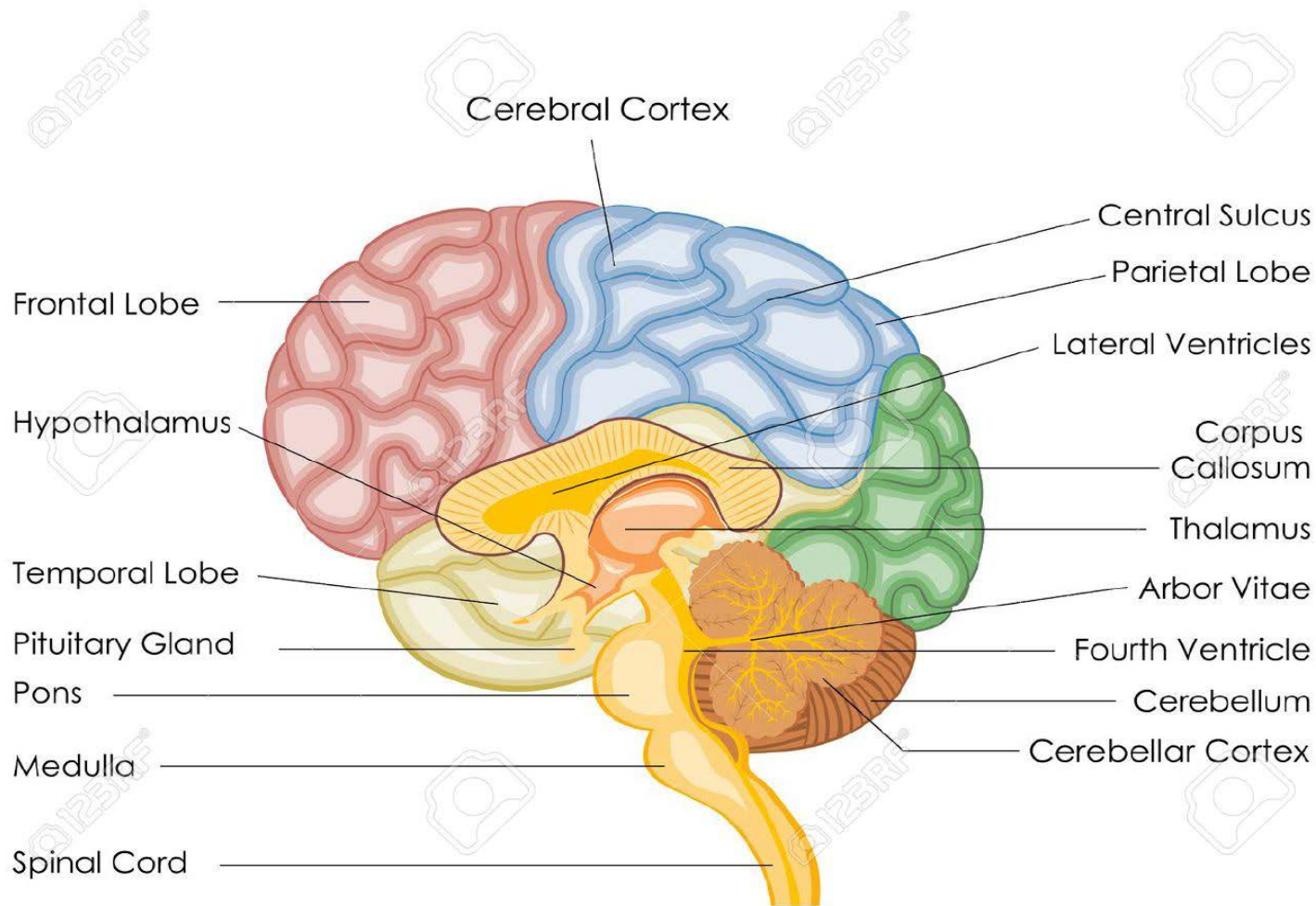


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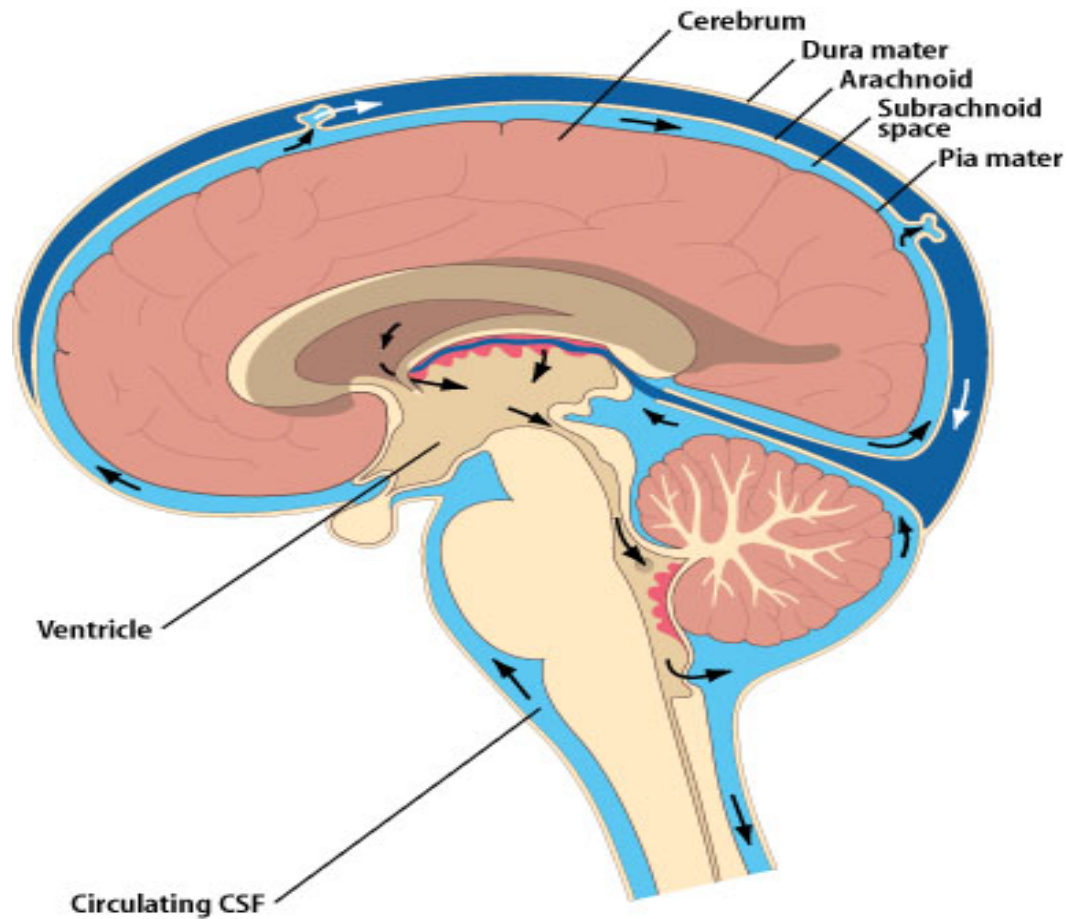
Non-Malignant CNS Equivalent Terms and Definitions
C700, C701, C709, C710-C719, C720-C725, C728, C729, C751-C753
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



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



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



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

Note 1: **Timing** is **not used** to determine multiple primaries.

Note 2: **Laterality** is **not used** to determine multiple primaries.

Note 3: **Malignant** central nervous system (CNS) tumors have a separate set of rules.

Note 4: 2007 MPH Rules and 2018 Solid Tumor Rules are used based on **date of diagnosis**.

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

Unknown if Single or Multiple Tumors

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

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

Note 2: Examples of cases with minimal information include

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

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

ⁱPrepare one abstract. Use the [histology rules](#) to assign the appropriate histology code.

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

Single Tumor

IMPORTANT: The major difference between **M3** and **M5** is:

M3: No resection as first course of treatment **AND** when the same tumor is subsequently **resected, pathology proves malignant** behavior

M5: Tumor resected as first course of treatment. Subsequent tumor (**recurrence** or **de novo**) is **malignant**

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

Note 1: A single tumor is **always** a single primary.

Note 2: The tumor may overlap onto or extend into an adjacent/contiguous site or subsite.

Note 3: The tumor may have multiple histologic components.

Note 4: The tumor may be diagnosed as borderline /1 on scans and subsequent pathology proves a benign /0 tumor.

Example: Patient has a MRI with a diagnosis of a benign /0 tumor. The pathology report from the subsequent surgery shows a borderline /1 tumor. This is a single tumor and a **single primary**. (It is the same tumor as seen on the MRI; better information is provided by the pathology report.)

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

1. The original diagnosis was non-malignant /0 or /1 **AND**

- First course treatment was active surveillance (no tumor resection). Diagnosis was:
 - o Clinical
 - o Radiographic
 - o 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**ⁱⁱ when a **malignant** tumor /3 occurs **after** a **non-malignant** tumor /0 or /1 **AND:**

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

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

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

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

Note 1: The bilateral tumors may appear simultaneously (at the same time) **OR** the contralateral tumor may be diagnosed at any time following the original diagnosis.

Note 2: WHO and IARC designate pilocytic astrocytoma as a synonym for optic glioma. When the primary site is optic nerve, the behavior is non-malignant.

Note 3: When the bilateral tumors are diagnosed at different times, the physician **may stage each tumor** because staging and determining multiple primaries are done for different reasons. Staging determines which course of treatment would be most effective. Determining multiple primaries is done to stabilize the data for the study of epidemiology (long-term studies done on incidence, mortality, and causation of a disease with the goal of reducing or eliminating that disease).

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

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

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

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

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

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

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

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

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

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

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

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

Note 1: Metastases are never used to determine multiple primaries. Seeding metastasis is often noted in ependymomas.

Note 2: This is a **change** from/**clarification** to **previous** rules.

Note 3: These rules are hierarchical. Use this rule **ONLY** when the previous rules do not apply.

Note 4: An example of a non-malignant brain tumor that may be multi-focal/multi-centric is hemangioblastoma **9161/1**.

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

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

Note: The same row means the tumors are:

- The same histology (same four-digit ICD-O code) **OR**
- One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
- A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3). NOS and subtype/variants are:
 - Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
 - Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
 - Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
 - Lipoma **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**ⁱⁱ when separate/non-contiguous tumors are on **different rows** in **Table 6** in the Equivalent Terms and Definitions. Timing is irrelevant.
Note: Each row in the table is a **distinctly different** histology.

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

This is the end of instructions for Multiple Tumors

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

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

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

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

Note 2: **Malignant** central nervous system (CNS) tumors have a **separate** set of **rules**.

Note 3: These rules are **not used** for tumor(s) or neoplasm(s) described as **metastatic/metastasis**.

Note 4: For rules specifying a **NOS** and a **subtype/variant** of the NOS, the NOS may be the preferred/most common term **OR** any of the **synonyms** for the NOS.

Priority Order for Using Documentation to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed prior to neoadjuvant treatment.
Note 1: Histology changes 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/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)

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 meningotheial meningioma. The treatment plan says the patient will receive the following treatment for meningotheial meningioma. Treatment plan confirms meningotheial meningioma; code meningotheial meningioma. The case meets the criteria in bullet 2.

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

List of Ambiguous Terminology

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

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

- Architecture
- Foci; focus; focal
- Pattern

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

Single Tumor

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

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

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

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

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

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

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

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

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

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

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

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

Rule H3 Code the histology when only **one histology** is present.
Note 1: Use [Table 6](#) to code histology. New codes, terms, and synonyms are included in **Table 6** and coding errors may occur if the table is not used.
Note 2: When the histology is **not listed** in **Table 6** use the **ICD-O** and all **updates**.
Note 3: Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in Table 6, ICD-O or all updates.

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

- Choroid plexus papilloma **9390/0** and a subtype/variant of choroid plexus papilloma
- Craniopharyngioma **9350/1** and a subtype/variant of craniopharyngioma
- Gangliocytoma **9492/0** and a subtype/variant of gangliocytoma
- Lipoma **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** /3 meningioma based on tumor extension/tumor expansion such as:

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

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

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

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

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

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

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

Note 1: Only report tumors such as:

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

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

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

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)

Introduction

Note 1: The group name “urinary sites” include: Renal pelvis C659; ureter C669; trigone of bladder C670; dome of bladder C671; lateral wall of bladder C672; anterior wall of bladder C673; posterior wall of bladder C674; bladder neck C675; ureteric orifice C676; urachus C677; overlapping lesion of bladder C678; bladder NOS C679; urethra C680; paraurethral gland C681; overlapping lesion of urinary organs C688; and urinary system NOS C689.

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

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

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

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

In US, 90% of bladder tumors are urothelial carcinoma; less than 5% are pure squamous cell carcinoma or pure adenocarcinoma.

Urothelial carcinoma originates in urothelial/transitional cells which line the urethra, bladder, ureters, and renal pelvis and has two major subdivisions: papillary and non-papillary.

- Papillary carcinoma: (commonly in bladder, ureter, or renal pelvis): A warty growth which projects from the wall on a stalk
 - Non-invasive papillary urothelial carcinoma (occasionally called in situ)
 - Invasive papillary urothelial carcinoma
- Non-papillary urothelial: originates within the mucosa and does not project from the wall
 - Non-invasive carcinoma in situ (CIS)
 - Invasive urothelial carcinoma

Note: Both urothelial carcinoma and papillary urothelial carcinoma can be in situ /2 or invasive /3. Code the behavior specified in the pathology report.

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
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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.

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Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
Note: “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**. 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
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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
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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 1: ICD-O Primary Site Codes

Use the following table to determine the correct site code.

Column 1 contains the site term and ICD-O code.

Column 2 contains synonyms for the site code and term in column 1.

Site Term and code	Synonyms
Bladder, anterior wall C673	-
Bladder, dome C671	Roof Vault Vertex
Bladder, lateral wall C672	Lateral to ureteral orifice Left wall Right wall Sidewall
Bladder neck C675	Internal urethral orifice Vesical neck
Bladder NOS C679	Lateral posterior wall (no hyphen)
Bladder, overlapping lesion C678	Fundus Lateral-posterior wall (hyphen)
Bladder, posterior wall C674	-

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
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Site Term and code	Synonyms
Bladder, trigone C670	Base of bladder Below interureteric crest Below interureteric field Below interureteric ridge Floor of bladder
Bladder, urachus C677	Mid umbilical ligament
Bladder, ureteric orifice C676	Just above ureteric orifice
Overlapping lesion of urinary organs C688	-
Paraurethral gland C681	-
Renal pelvis C659	Pelvis of kidney Pelviureteric junction Renal calyces Renal calyx
Ureter C669	-
Urethra C680	Cowper gland Prostatic utricle Urethral gland
Urinary system NOS C689	-

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
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Table 2: Specific Histologies, NOS, and Subtypes/Variants

Use Table 2 as directed by the [Histology Rules](#) to assign the more common histology codes for urinary tract neoplasms.

Column 1 contains specific and NOS histology terms.

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

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

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

Column 3 may contain NOS histologies which are part of a bigger histologic group. For example, sarcoma NOS 8800/3 (column 1) is a generic term which encompasses a number of soft tissue tumors, including rhabdomyosarcoma 8900/3 (column 3).

Rhabdomyosarcoma is also a NOS because it has a subtype/variant 8910/3. The subtype/variant is indented under the NOS (rhabdomyosarcoma) in column 3. There is also a note in column 1 which calls attention to the fact that rhabdomyosarcoma has a subtype/variant.

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

Table begins on next page

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

Specific and NOS Histology Codes	Synonyms	Subtypes/Variants
Adenocarcinoma NOS 8140 <i>Note:</i> Urachal carcinoma NOS is coded 8010/3. Urachal adenocarcinoma is coded 8140/3.	Mixed adenocarcinoma Urachal adenocarcinoma	Clear cell carcinoma 8310 Endometrioid carcinoma 8380 Enteric adenocarcinoma 8144 Mucinous adenocarcinoma 8480
Malignant melanoma 8720/3		
Malignant perivascular epithelioid cell tumor 8714/3	Malignant PEComa	
Sarcoma NOS 8800/3 <i>Note:</i> Rhabdomyosarcoma 8900 is a NOS with a subtype/variant of embryonal rhabdomyosarcoma/sarcoma botryoides 8910/3 .		Angiosarcoma 9120/3 Chondrosarcoma 9220/3 Leiomyosarcoma 8890/3 Liposarcoma 8850/3 Malignant peripheral nerve sheath tumor (MPNST) 9540/3 Pleomorphic sarcoma 8802/3 Rhabdomyosarcoma 8900/3 Embryonal rhabdomyosarcoma/sarcoma botryoides 8910/3
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

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

Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
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Table 3: Non-Reportable Urinary Tumors

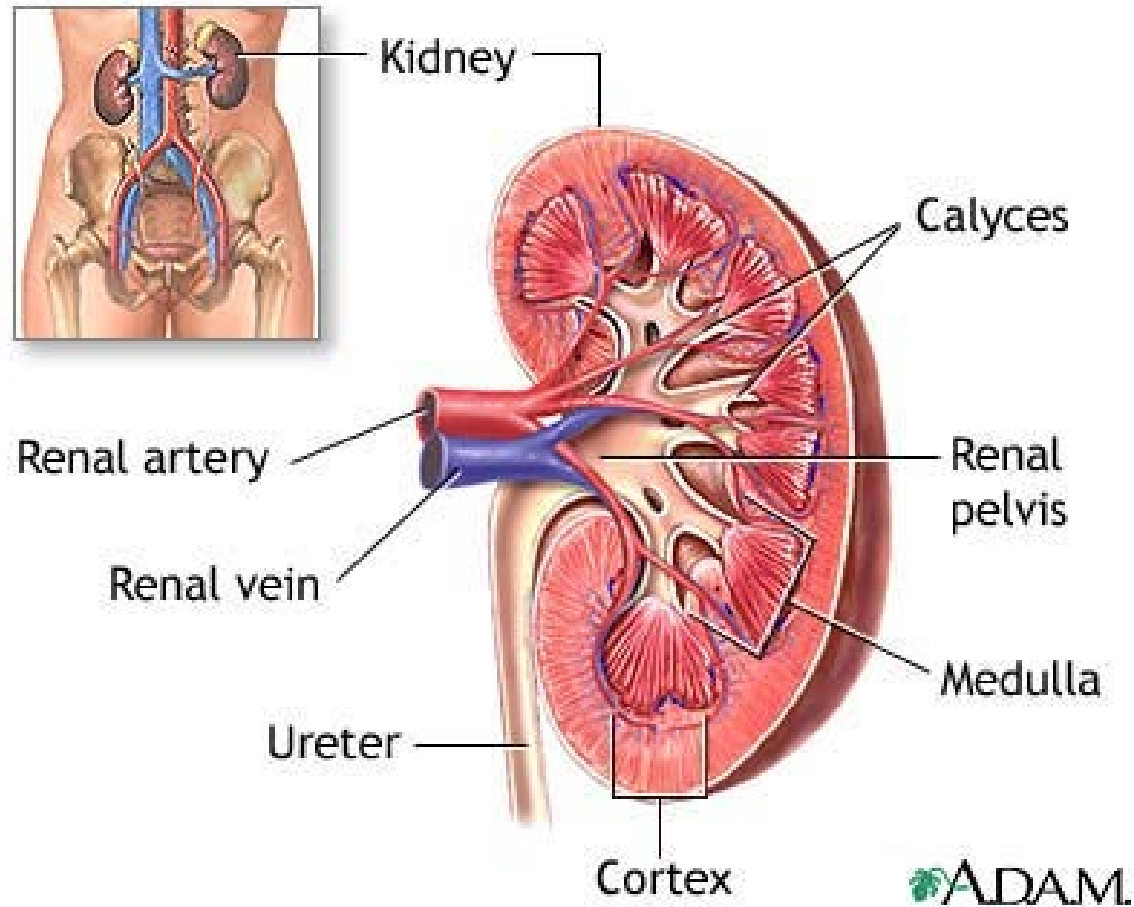
Column 1 contains the terms and codes (if applicable) for the non-reportable histology.

Column 2 contains **synonyms** of the histology term in column 1. Synonyms have the **same code** as the term in Column 1.

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

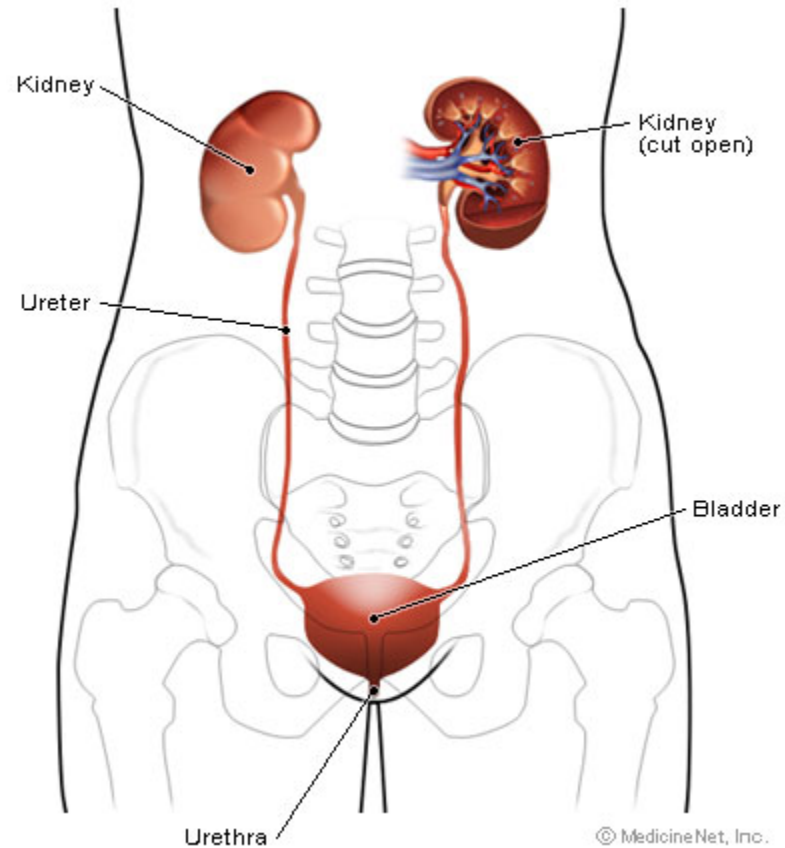
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Illustrations



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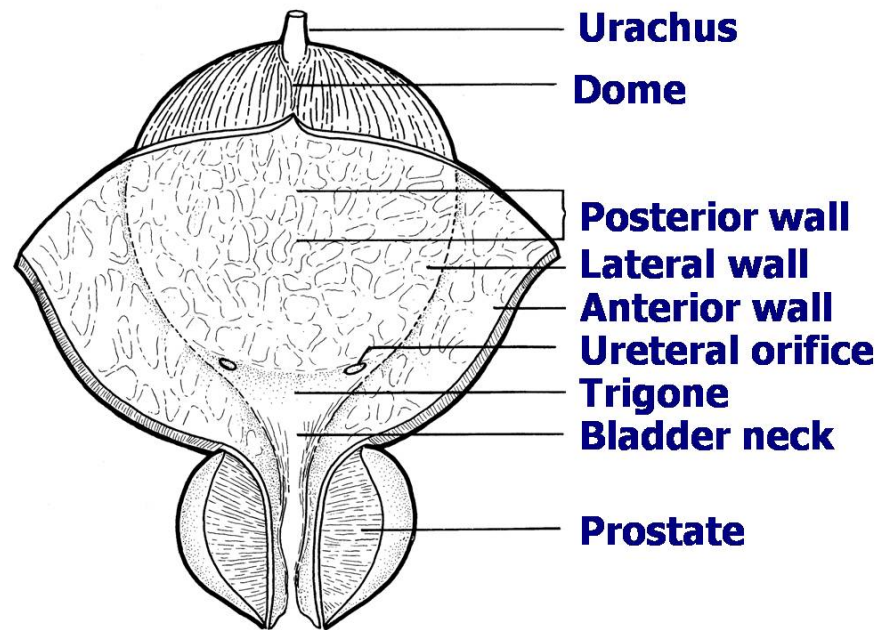
Renal Pelvis, Ureter, Bladder, and Other Urinary Equivalent Terms and Definitions
C659, C669, C670-C679, C680-C689
(Excludes lymphoma and leukemia M9590 – M9992 and Kaposi sarcoma M9140)



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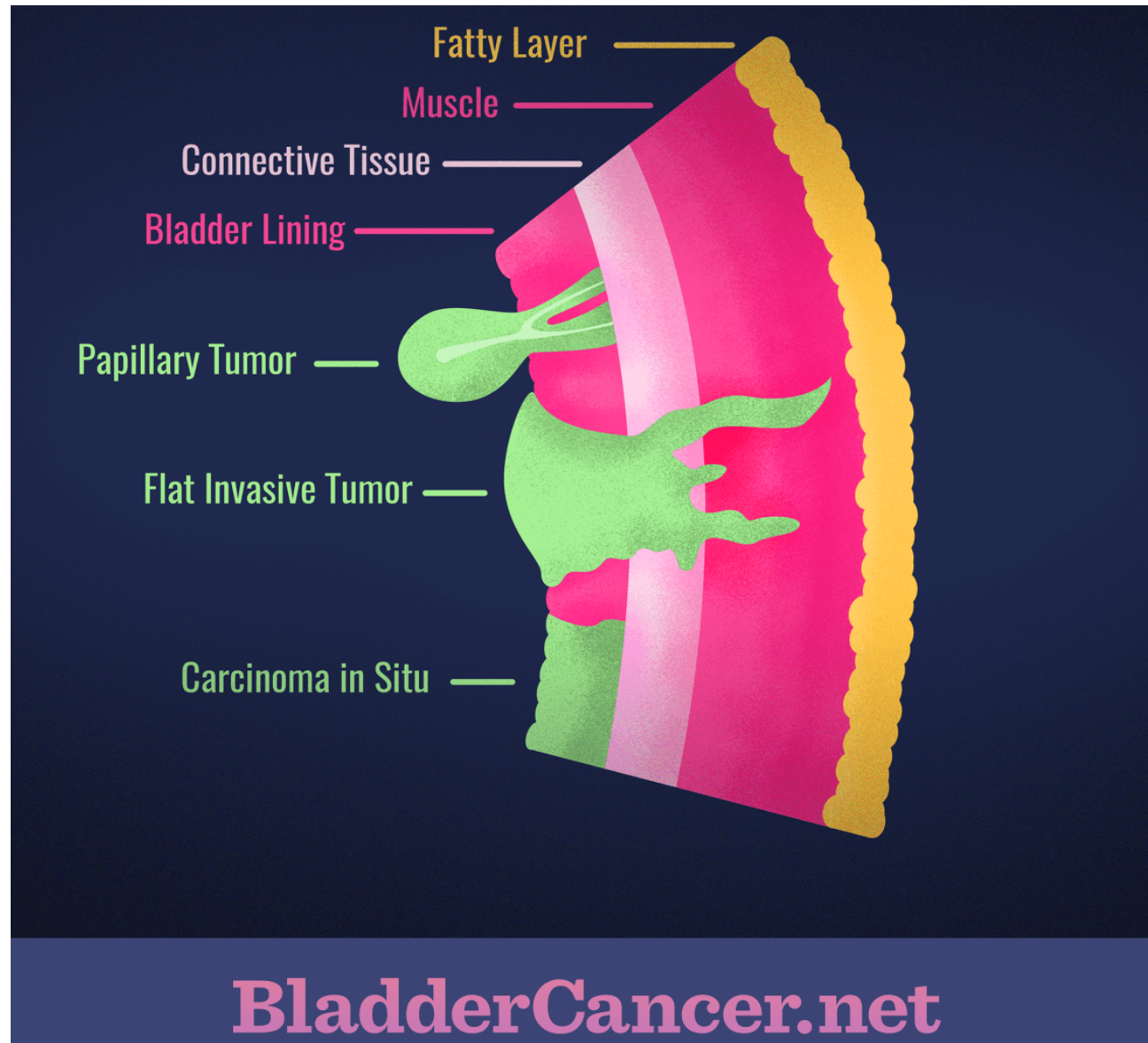
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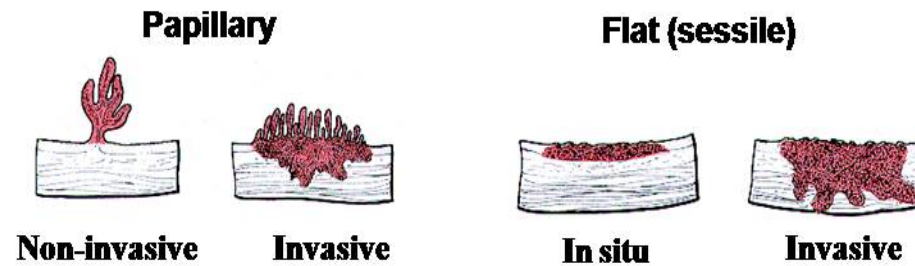


Source: TNM Atlas, 3rd edition, 2nd revision

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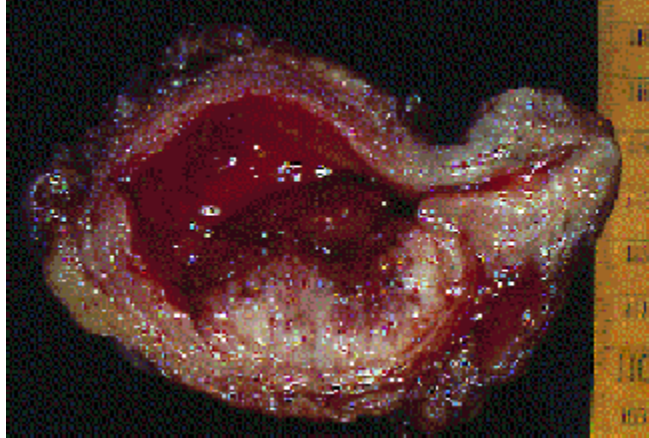


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Bladder Tumor

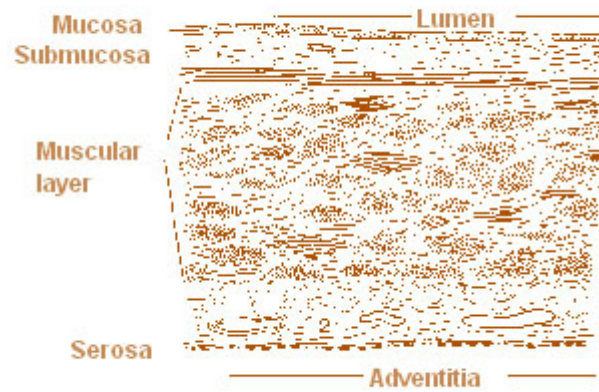


Courtesy Jean-Loup Huret reprinted from Atlas Genet Cytogenet Oncol Haematol. October 2003. van Tilborg A

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

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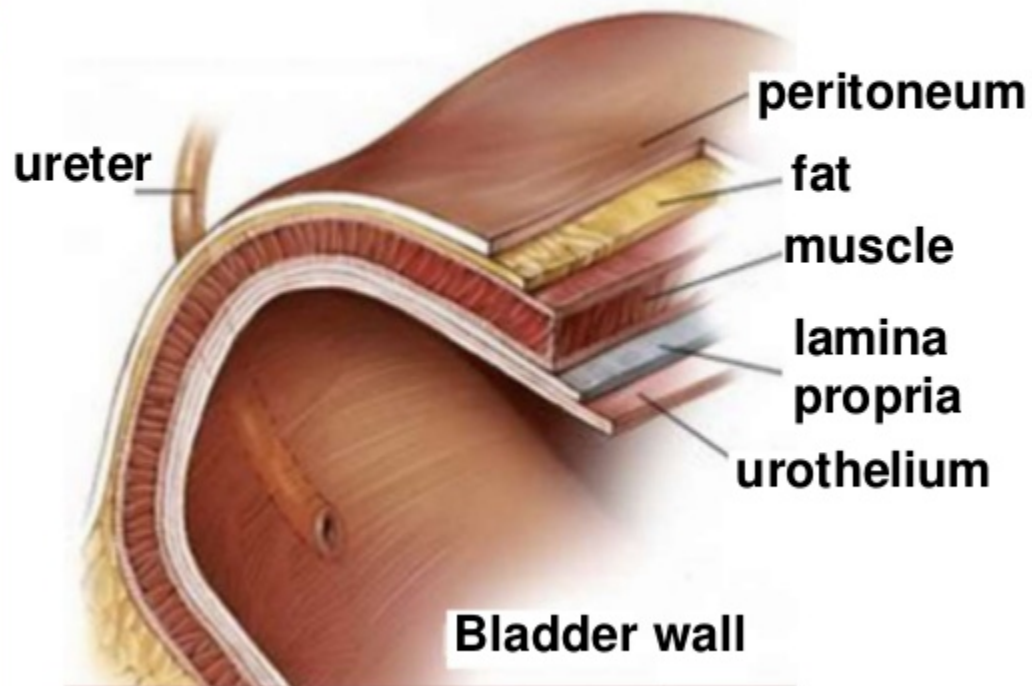
Bladder Wall



Source: Feneis, Pocket Atlas of Human Anatomy, 2nd ed.

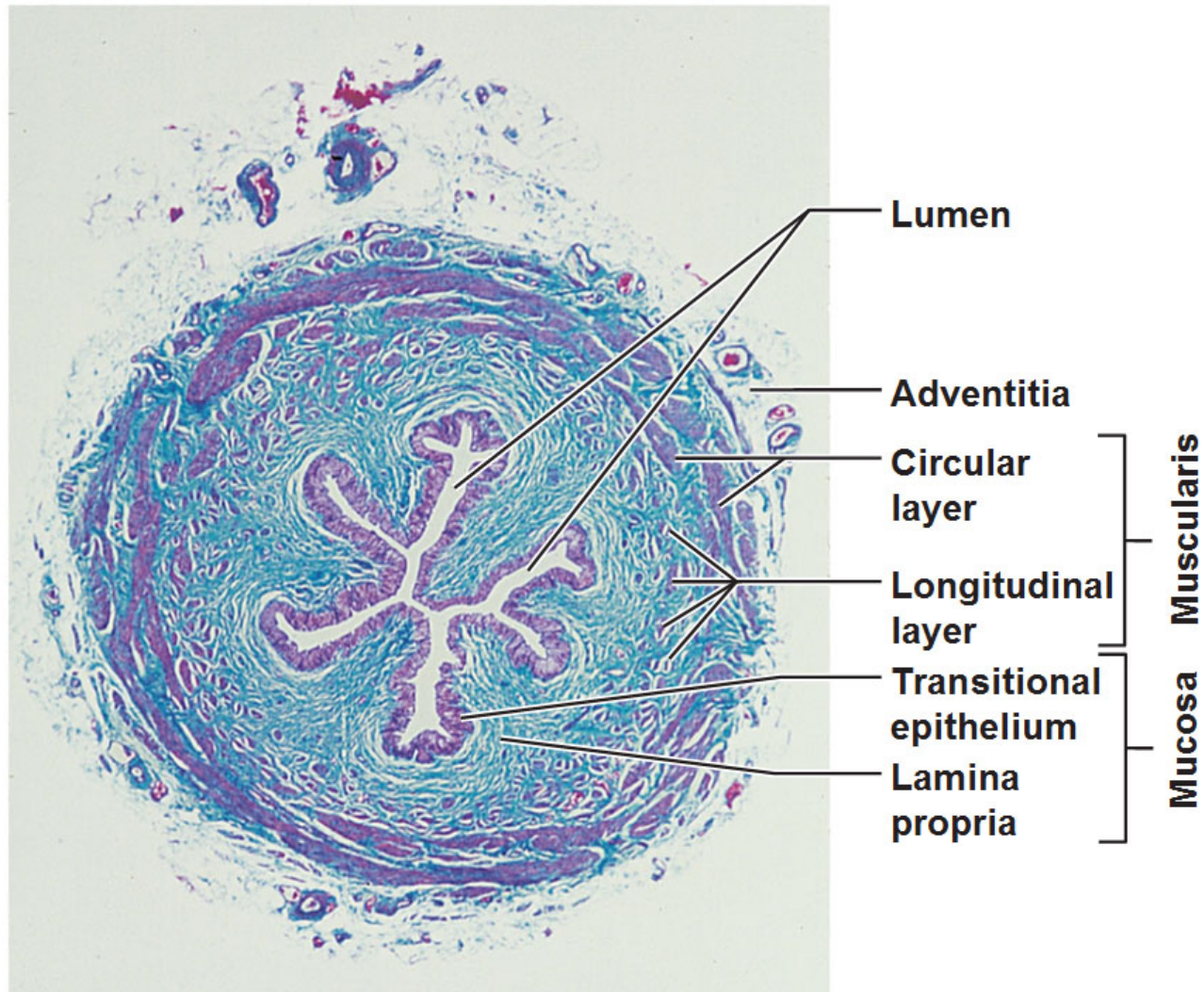
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Layers of the Bladder Wall



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Microscopic Structure of the Ureter



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

- Bones
- Brain
- Regional and distant lymph nodes for the primary site being abstracted as identified in Summary Staging Manual
- Involvement of the pelvic or abdominal wall
- Liver
- Lung

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

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

Unknown if Single or Multiple Tumors

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

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

Note 2: Examples of cases with minimal information include:

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

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

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

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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.

ⁱPrepare one abstract. Use the [histology rules](#) to assign the appropriate histology code.

Multiple Tumors

Note 1: Multiple tumors may be a single primary or multiple primaries.

Note 2: Separate, non-contiguous tumors are always multiple primaries when:

- In the urinary system (see [Table 1](#)) **AND** in a site other than the urinary system
Example: Patient has urothelial carcinoma of the bladder and non-metastatic adenocarcinoma of the lung. The lung is not a urinary site. Abstract two primaries.
- Non-synchronous tumors other than urothelial carcinoma and urothelial carcinoma subtypes in multiple urinary sites (see Rule M14)

Rule M3 Abstract **multiple primaries**ⁱⁱ when there are:

- Separate/non-contiguous tumors in both the right **AND** left **renal pelvis AND**
- No other urinary sites are involved with separate/non-contiguous tumors

Note 1: Only abstract a single primary when pathology confirms tumor(s) in the contralateral renal pelvis are metastatic.

Note 2: This rule is used only when there is no involvement by separate/non-contiguous tumors in the ureter(s), bladder, or urethra.

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Rule M4 Abstract **multiple primaries**ⁱⁱ when there are:

- Separate/non-contiguous tumors in the right **AND** left **ureter AND**
- No other urinary sites are involved with separate/non-contiguous tumors

Note 1: Only abstract a single primary when pathology confirms tumor(s) in contralateral ureter are metastatic.

Note 2: This rule is used **only** when there is **no involvement** by separate/non-contiguous tumors in the renal pelvis, bladder, and urethra.

Rule M5 Abstract a **single primary**ⁱ when **synchronous** tumors are **noninvasive in situ /2** urothelial carcinoma (flat tumor) **8120/2 in the following sites:**

- **Bladder C67_ AND**
- One or both **ureter(s) C669**

Note 1: No other urinary organs are involved.

Note 2: Use this rule **ONLY** for noninvasive in situ urothelial carcinoma (may be called noninvasive urothelial carcinoma or noninvasive flat tumor). For other histologies, continue through the rules.

Note 3: Urothelial carcinoma in situ spreads by intramucosal extension and may involve large areas of mucosal surface. The default for these cases is coding a bladder primary.

Rule M6 Abstract **multiple primaries**ⁱⁱ when an **invasive** tumor occurs **more than 60** days after an **in situ** tumor.

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

Note 2: Abstract as multiple primaries even if physician states the invasive tumor is disease recurrence or progression.

Note 3: This rule is based on long-term epidemiologic studies of recurrence intervals. The specialty medical experts (SMEs) reviewed and approved these rules. Many of the SMEs were also authors, co-authors, or editors of the AJCC Staging

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Rule M7 Abstract a **single primary**ⁱ when the patient has multiple occurrences of /2 urothelial carcinoma in the **bladder**. Tumors may be any combination of:

- In situ urothelial carcinoma **8120/2 AND/OR**
- Papillary urothelial carcinoma noninvasive **8130/2** (does **not** include micropapillary subtype)

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

Note 2: Abstract only one /2 urothelial bladder primary per the patient's lifetime.

Note 3: There are no /2 subtypes for urothelial carcinoma with the exception of papillary urothelial carcinoma.

Example: On 1/3/2018, the patient had a TURB with a diagnosis of in situ urothelial carcinoma 8120/2. On 5/8/2019, pathology from TURB is papillary urothelial carcinoma non-invasive 8130/2. This is a single primary; the papillary urothelial carcinoma is recorded as a recurrence for those registrars who collect recurrence data.

Rule M8 Abstract **multiple primaries**ⁱⁱ when the patient has **micropapillary** urothelial carcinoma **8131/3 of the bladder** **AND** a urothelial carcinoma **8120/3** (including papillary **8130/3**) **of the bladder**.

Note 1: This is a new rule for 2018.

Note 2: Micropapillary urothelial cell carcinoma is an extremely aggressive neoplasm. It is important to abstract a new primary to capture the incidence of micropapillary urothelial carcinoma. Micropapillary is excluded from the typical "NOS and subtype/variant" rule (same row in Table 2).

Rule M9 Abstract a **single primary**ⁱ when the patient has multiple **invasive** urothelial cell carcinomas in the **bladder**. All tumors are **either**:

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

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

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

- An occurrence of micropapillary and an occurrence of urothelial carcinoma would be multiple primaries (see previous rules).

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

Note 1: This rule **does not apply** when both/all tumors are urothelial carcinoma of the bladder.

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

- Scans are NED
- Urine cytology is NED
- Scopes are NED

Note 3: When there is a recurrence within three years of diagnosis, the “**clock**” starts over. The time interval is calculated from the **date of last recurrence**.

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

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

Example: Patient is diagnosed with multifocal/multicentric urothelial carcinomas in the ureter and renal pelvis in January 2018. Both the kidney and ureter are surgically removed. In June 2022 the patient presents with tumor in the contralateral ureter. The physician states this is a recurrence of the original urothelial carcinoma. Code a new primary for the 2022 ureter carcinoma.

Rule M11 Abstract a **single primary**ⁱ when there are **urothelial carcinomas** in multiple urinary organs.

Note 1: This rule is **ONLY** for urothelial carcinoma **8120** and all subtypes/variants of urothelial carcinoma. This rule does not apply to any other carcinomas or sarcomas.

Note 2: Behavior is irrelevant.

Note 3: This rule applies to multifocal/multicentric carcinoma which involves two or more of the following urinary sites:

- Renal pelvis
- Ureter
- Bladder
- Urethra

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Rule M12 Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are two or more **different subtypes/variants** in Column 3 of [Table 2](#) in the Equivalent Terms and Definitions. Timing is irrelevant.

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

- **Same NOS:** Leiomyosarcoma 8890/3 and liposarcoma 8850/3 are both subtypes of sarcoma NOS 8800/3 but are distinctly different histologies. Abstract multiple primaries.
- **Different NOS:** Verrucous carcinoma 8051 is a subtype of squamous cell carcinoma NOS 8070; giant cell urothelial carcinoma 8031 is a subtype of urothelial carcinoma 8120. They are distinctly different histologies. Abstract multiple primaries.

Rule M13 Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are on **different rows** in [Table 2](#) in the Equivalent Terms and Definitions. Timing is irrelevant.

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

Example: Small cell neuroendocrine carcinoma 8041 and urothelial carcinoma 8120 are on different rows of Table 2. Abstract two primaries, one for the small cell neuroendocrine carcinoma and a second for the urothelial carcinoma.

Rule M14 Abstract **multiple primaries**ⁱⁱ when the ICD-O **site** code differs at the **second** (C**X**xx) and/or **third** (Cx**X**x) character.

Rule M15 Abstract a **single primary**ⁱ when **synchronous**, separate/non-contiguous tumors are on the **same row** in [Table 2](#) in the Equivalent Terms and Definitions.

Note: The same row means the tumors are:

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

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.

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Rule M16 Abstract a **single primary**ⁱ (the invasive) when an **in situ** tumor is diagnosed **after** an **invasive** tumor **AND** tumors occur in the **same** urinary site.

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

Note 2: The tumors may be a NOS and a subtype/variant of that NOS. See [Table 2](#) in the Equivalent Terms and Definitions for listings of NOS and subtype/variants.

Note 3: Once the patient has an invasive tumor, the subsequent **in situ** is recorded as a **recurrence** for those registrars who collect recurrence data.

Rule M17 Abstract a **single primary**ⁱ (the invasive) when an invasive tumor is diagnosed **less than or equal to 60 days after** an **in situ** tumor **AND** tumors occur in the **same** urinary site.

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

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

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

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

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

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

Rule M18 Abstract a **single primary**ⁱ when tumors do not meet any of the above criteria.

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

This is the end of instructions for Multiple Tumors.

ⁱPrepare 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.

ⁱⁱPrepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.

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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 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

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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.

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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.

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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 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.

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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
- Configuration
- Foci; focus; focal
- Pattern

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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/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.

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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.

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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.

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 H9** Code mixed small cell carcinoma **8045** when the final diagnosis **for all tumors** is any of the following:
- Small cell neuroendocrine mixed with any other type of **carcinoma** (does not apply to sarcoma)
 - Two or more subtypes/variants of small cell neuroendocrine carcinoma
 - Subtype/variant of small cell neuroendocrine mixed with any other carcinoma (does not apply to sarcoma)
- Example:* Diagnosis from TURB is urothelial carcinoma **and** small cell neuroendocrine carcinoma. Code mixed small cell carcinoma 8045.

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

Code the histology using the rule that fits the case.

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Introduction

- Note 1:** Tables and rules refer to ICD-O rather than ICD-O-3. The version is not specified to allow for updates. Use the currently approved version of ICD-O.
- Note 2:** 2007 MPH Rules and 2021 Solid Tumor Rules are used based on date of diagnosis
- Tumors diagnosed 01/01/2007 through 12/31/2020: Use 2007 MPH Rules and 2007 General Instructions
 - Tumors diagnosed 01/01/2021 and later: Use 2021 Solid Tumor Rules and Solid Tumor General Instructions
 - The original tumor diagnosed *before* 01/01/2021 and a subsequent tumor diagnosed 01/01/2021 or later in the same primary site: Use the 2021 Solid Tumor Rules and Solid Tumor General Instructions
- Note 3:** Melanoma can also start in the mucous membranes of the mouth, anus and vagina, in the eye or other places in the body where melanocytes are found. This scheme is used *only* for melanomas that *occur on the skin*.
- Note 4:** The WHO Classification of Skin Tumors 4th Ed does not include ICD-O codes for tumors with mixed melanoma subtypes/variants
- Note 5:** Cutaneous melanoma starts in the melanocytes of the skin. Melanocytes lie in the epidermis, the outermost layer of the skin. Melanocytes often cluster together and form moles (nevi). Most moles are benign, but some may become malignant melanomas. Melanomas are divided into 5 main types, depending on their location, shape, and whether they grow outward or downward into the dermis:
- Acral melanoma: occurs on the palms of the hand, soles of the feet, or nail beds
 - Desmoplastic melanoma: is a rare malignant melanoma marked by non-pigmented lesions on sun exposed areas of the body
 - Lentigo maligna: usually occur on the faces of elderly people
 - Superficial spreading or flat melanoma: grows outwards at first to form an irregular pattern on the skin with an uneven color
 - Nodular melanoma: are lumpy and often blue-black in color and may grow faster and spread downwards

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Changes from 2007 MPH Rules

These changes are effective with cases diagnosed 1/1/2021 and later. WHO 4th Ed Classification of Skin Tumors was published in 2018.

1. 2007 Rules instruct “Code the histology from the most representative specimen.” For all sites except breast and CNS, the 2021 Rules instruct “Code the most specific histology from biopsy or resections”. When there is a discrepancy between the biopsy and resection (two distinctly different histologies), code the histology from the most representative specimen (the greater amount of tumor).” **This instruction applies to the 2021 cutaneous melanoma solid tumor rules.**
2. Early/evolving melanoma in situ (8720/2) and early/evolving melanoma invasive (8720/3) are reportable for cases diagnosed 1/1/2021 and later. Please refer to [SEER Program Coding and Staging Manual 2021](#) for additional information on reportable neoplasms.
3. New histology **terms** are included (identified by asterisks (*) in the histology table in the Terms and Definitions). No new cutaneous melanoma ICD-O histology **codes** have been proposed by WHO.
4. Some histologies are rare and may not be listed in the tables; refer to ICD-O and all updates. If the histology is not found in the tables or ICD-O, submit a question to [Ask a SEER Registrar](#).
5. WHO 4th Ed Skin Tumors now classifies melanocytic tumors into two groups:
 - A. Melanomas arising in sun-exposed skin
 - B. Melanomas arising at sun-shielded sites or without known etiological association with UV radiation exposure

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Equivalent or Equal Terms

These terms can be used interchangeably:

- And; with
Note: “And” and “with” are used as synonyms when describing multiple histologies within a single tumor.
- Giant pigmented nevus; giant congenital nevus
- Mixed epithelioid and spindle cell melanoma (8770); Epithelioid melanoma and spindle cell melanoma
- Melanoma in situ, superficial spreading type; low-cumulative sun damage (CSD) melanoma in situ
- Mole; Nevus
- Simultaneous; existing at the same time; concurrent
- Site; topography
- Superficial spreading melanoma; low-cumulative sun damage (CSD) melanoma
- Tumor; mass; tumor mass; lesion; neoplasm
 - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a **standard manner** in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician’s statement that the term is malignant/melanoma**
 - These terms are used **ONLY** to determine multiple primaries
 - **Do not** use these terms for casefinding or determining reportability
- Type; subtype; variant

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terms that are not Equivalent or Equal

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

- Component is not equivalent to subtype/type/variant
Note 1: Component is only coded when the pathologist specifies the component as a second *melanoma*
Note 2: Examples provided in H rules [Coding Histology](#) section
- Phenotype is not equivalent to subtype/type/variant

Synonyms for In Situ

- Behavior code 2
- Clark level 1 (limited to the epithelium)
- Hutchinson freckle (See [Synonyms for Hutchinson Freckle](#))
- Intraepidermal, NOS
- Intraepithelial, NOS
- Lentigo maligna
- Noninvasive
- Precancerous melanoma of Dubreuilh
- Precancerous melanosis (C44_)
- Stage 0
- Tis

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Synonyms for Hutchinson Freckle

- Circumscribed precancerous melanosis
- Intraepidermal malignant melanoma
- Lentigo maligna
- Precancerous melanosis of Dubreuilh

Anatomical Dermatology Terms

Term	Definition
Cutaneous	Pertaining to skin
Dermal	Pertaining to skin
Epidermal	Pertaining to upon the skin
Hypodermic	Pertaining to below the skin
Intradermal	Pertaining to within the skin
Subcutaneous	Pertaining to under the skin
Ungual	Pertaining to the nail

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Table 1: Primary Sites and Laterality

Table 1 contains terms used in **clinical diagnosis**, and less frequently the **operative and pathology reports** to describe the **location** of the skin lesion. Find the **term** in Column 1 and use the **site code** in Column 2. Column 3 notes whether the site requires **laterality** to be coded.

Note: Excludes melanoma of non-skin sites (excludes any sites other than C44_)

Terminology	Site Term and Code	Laterality Required
Skin of lip, NOS Skin of lower lip Skin of upper lip	Skin of lip, NOS C440	No
Eyelid Lid, NOS Palpebra Horizontal palpebra fissure Canthus Inner canthus Lateral canthus Lower lid Medial canthus Meibomian gland Outer canthus Pretarsal space Subpratarsel crease Upper lid	Eyelid C441	Yes

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
External ear Auricle, NOS Pinna Ceruminal gland Concha Ear, NOS Ear lobule Earlobe External auditory canal Auditory canal, NOS Auricular canal, NOS External auricular canal Ear canal External auditory meatus Helix Skin of auricle Skin of ear, NOS Tragus	External ear C442	Yes

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of other & unspecified parts of face Skin of: Cheek Chin Face Forehead Jaw Nose Temple Ala nasi Chin, NOS Columnella Eyebrow Brow External cheek External nose Forehead, NOS Lid-cheek junction Nasaljugal groove Temple, NOS	Skin of other and unspecified parts of face C443	Yes
Skin of scalp and neck Skin of head, NOS Skin of neck Skin of scalp Scalp, NOS Skin of cervical region	Skin of scalp and neck C444	Yes (New)

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of trunk Skin of: Abdomen Abdominal wall Anus Axilla Back Breast Buttock Chest Chest wall Flank Groin Perineum Thoracic wall Thorax Trunk Umbilicus Gluteal region Infraclavicular region Inguinal region Sacroccocygeal region Scapular region Perianal skin Umbilicus, NOS	Skin of trunk C445	Yes

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
Skin of upper limb and shoulder Skin of: Antecubital space Arm Elbow Finger Forearm Hand Palm Shoulder Thumb Upper limb Wrist Fingernail Nail Bed Palmar skin	Skin of upper limb and shoulder C446	Yes

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Terminology	Site Term and Code	Laterality Required
<p>Skin of lower limb and hip Skin of: Ankle Calf Foot Heel Hip Knee Leg Lower limb Popliteal space Thigh Toe Plantar skin Sole of foot Toenail</p>	<p>Skin of lower limb and hip C447</p>	<p>Yes</p>
<p>Overlapping lesion of skin</p>	<p>Overlapping lesion of skin C448 For Head and Neck: Do not use C448 for overlapping lesions of the Head & Neck. Assign the primary site code for the site where the bulk of the tumor is or where the epicenter is; do not use code C448.</p>	<p>No</p>
<p>Skin, NOS <i>Note:</i> Code to Skin, NOS (C44.9) if a patient is diagnosed with metastatic melanoma and the primary site is not identified.</p>	<p>Skin, NOS C449</p>	<p>No</p>

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Table 2: Specific Histologies, NOS, and Subtype/Variants

Use Table 2 as directed by the [Histology Rules](#) to assign the more common histology codes for melanotic skin tumors

Note 1: Rare histologies may not be listed in the table. When a histology term is not found, reference ICD-O and all updates.

Note 2: Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in Table 2, ICD-O or ICD-O updates.

Note 3: Behavior codes are listed when the term has only one possible behavior (either a /2 or /3). For histologies which may be either /2 or /3, a behavior code is not listed. Code behavior as documented in the pathology report.

Note 4: Only use the histology code from the table when the diagnosis is **EXACTLY** the term listed.

Note 5: **New histology terms are identified by asterisk (*)**

Column 1 contains specific and NOS histology terms.

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

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

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

Table begins on the next page

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

****Terms approved by standard setters and are not listed in WHO or ICD-O**

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

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

NOS Histology Terms and Codes	Synonyms	Subtypes/Variants
<p>Melanoma, NOS 8720</p> <p><i>Note:</i> Sarcomatoid melanoma is a rare subtype of melanoma characterized by almost complete loss of melanocytic differentiation both morphologically and phenotypically, with the bulk of the tumor being replaced by a spindle cell, sarcomatoid component. Use code 8772/3, spindle cell melanoma.</p>	<p>Melanoma in situ 8720/2</p> <p>Early/Evolving melanoma in situ** 8720/2</p> <p>Nevoid melanoma 8720/3</p> <p>Early/Evolving invasive melanoma** 8720/3</p>	<p>Acral melanoma*/acral lentiginous melanoma, malignant 8744/3</p> <p>Amelanotic melanoma 8730/3</p> <p>Balloon cell melanoma 8722/3</p> <p>Desmoplastic melanoma/desmoplastic melanoma, amelanotic/neurotropic melanoma, malignant 8745/3*</p> <p>Epithelioid cell melanoma 8771/3</p> <p>Lentigo maligna/Hutchinson melanotic freckle 8742/2</p> <p>Lentigo maligna melanoma/Melanoma in Hutchinson melanotic freckle 8742/3</p> <p>Low cumulative sun damage melanoma*/superficial spreading melanoma 8743/3</p> <p>Melanoma arising in a blue nevus 8780/3*</p> <p>Malignant melanoma arising in giant congenital nevus*/malignant melanoma in giant pigmented nevus 8761/3</p> <p>Malignant melanoma in a precancerous melanosis 8741/3</p> <p>Malignant melanoma, regressing 8723/3</p> <p>Malignant Spitz tumor*/mixed epithelioid and spindle cell melanoma 8770/3</p> <p>Nodular melanoma 8721/3</p> <p>Spindle cell melanoma 8772/3</p> <p>Spindle cell melanoma, type A 8773/3</p> <p>Spindle cell melanoma, type B 8774/3</p>

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Table 3: Non-Reportable Neoplasms

Table 3 lists non-reportable terms and codes used in the diagnosis of cutaneous melanotic neoplasms. *This table is intended to be a reference only and may not be complete.* Please refer to your standard setter program manual for additional information on reportable neoplasms.

Non-Reportable Histology Term	Non-Reportable Histology Code
Pigmented nevus, NOS Nevus, NOS Melanocytic nevus Hairy nevus Nevus spilus Meyerson nevus Deep penetrating nevus Combined nevus Genital nevus Conjunctival nevus	8720/0
Balloon cell nevus	8722/0
Halo nevus Regressing nevus	8723/0
Neuronevus	8725/0
Magnocellular nevus Melanocytoma, NOS	8726/0
Dysplastic nevus	8727/0
Nonpigmented nevus Achromic nevus	8730/0
Junctional nevus, NOS Intraepidermal nevus Junction nevus	8740/0

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Non-Reportable Histology Term	Non-Reportable Histology Code
Lentiginous melanocytic nevus Simple lentigo Lentigo simplex	8742/0
Acral nevus	8744/0
Dermal nevus Intradermal nevus Stromal nevus	8750/0
Compound nevus Dermal and epidermal nevus	8760/0
Congenital melanocytic nevus, NOS Giant pigmented nevus, NOS Intermediate and giant congenital nevus	8761/0 8761/0 8761/1
Proliferative dermal lesion in congenital nevus Proliferative nodule in congenital melanocytic nevus	8762/1
Epithelioid and spindle cell nevus Juvenile melanoma Juvenile nevus Spitz nevus Spitz nevus, atypical Pigmented spindle cell nevus of Reed Pigmented spindle cell Spitz nevus	8770/0
Epithelioid cell nevus	8771/0
Spindle cell nevus, NOS	8772/0
Blue nevus, NOS Jadassohn blue nevus Pigmented epithelioid melanocytoma Blue nevus, epithelioid	8780/0 8780/0 8780/1 8780/1
Cellular blue nevus	8790/0

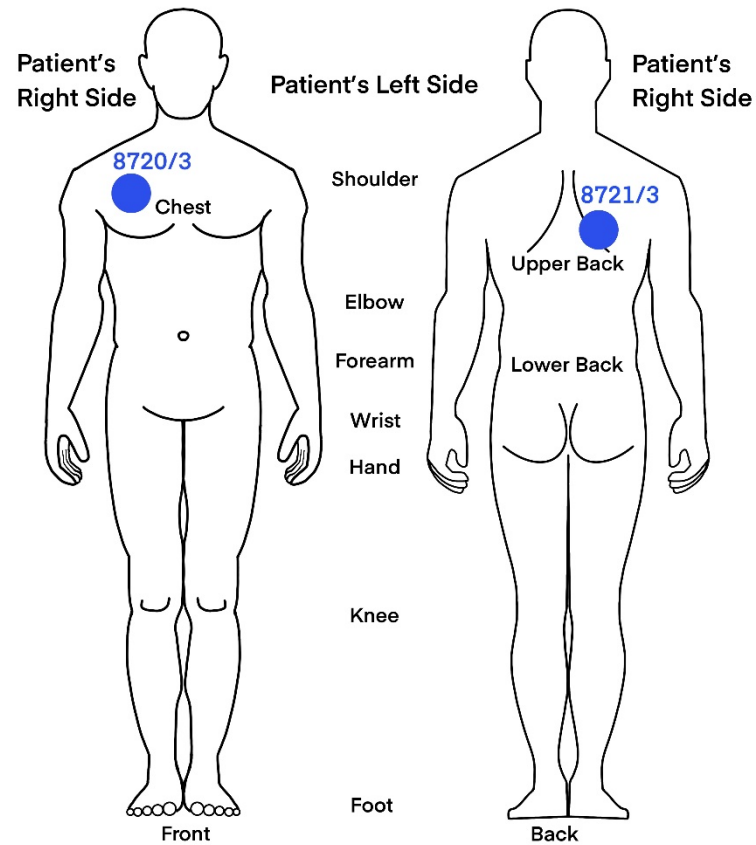
Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Non-Reportable Histology Term	Non-Reportable Histology Code
Intermediate lesion Melanocytic neoplasm of low malignant potential Melanocytic tumor of uncertain malignant potential (MELTUMP) Superficial atypical melanocytic proliferation of uncertain significance (SAMPUS) Primary acquired melanosis	No ICD-O code

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Illustrations

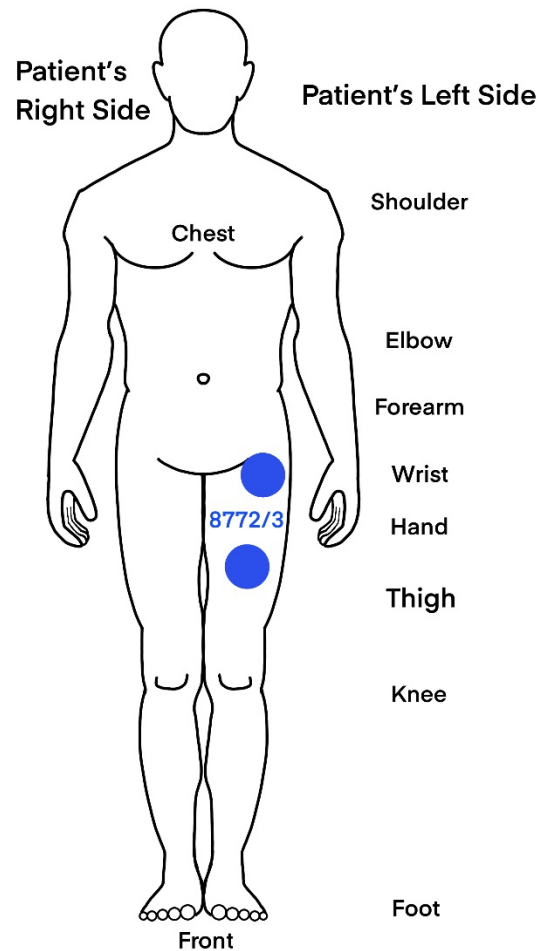
Explanatory illustrations for Multiple Primary Rule M6



Example 1: Both lesions are located on the right side of the body and sites are chest C445 and back C445. Histology for the chest lesion is melanoma, NOS (8720/3) and the back lesion is nodular melanoma (8721/3). Abstract a single primary.

Cutaneous Melanoma Equivalent Terms and Definitions
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Explanatory illustrations for Multiple Primary Rule M6



Example 2: Both lesions are located on the left leg. One lesion is spindle cell melanoma (8772/3) located on the front of the left hip C447. The other lesion is spindle cell melanoma (8772/3) located on the front of the left thigh C447.

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

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

- Distant metastasis in skin, subcutaneous tissue including muscle
- Bone
- Brain
- Regional lymph nodes as identified in Summary Stage Manual
- Distant lymph nodes as identified in Summary Stage Manual
- Liver
- Lung
- In-transit metastases – metastases which occur along the lymph pathways between the primary tumor > 2 cm from the scar and the regional lymph nodes
- Satellites – new tumor within a radius of 2 cm from the scar after removal of primary tumor. Satellites may be caused by remains of the primary tumor.

Note 2: 2007 MPH Rules and 2021 Cutaneous Melanoma Solid Tumor Rules are used based on **date of diagnosis**.

- Tumors diagnosed 01/01/2007 through 12/31/2020: Use 2007 MPH Rules and 2007 General Instructions
- Tumors diagnosed 01/01/2021 and later: Use 2021 Solid Tumor Rules
- The original tumor diagnosed **before** 1/1/2021 and a subsequent tumor diagnosed 1/1/2021 or later in the same primary site: Use the 2021 Solid Tumor Rules

Unknown if Single or Multiple Melanomas

Rule M1 Abstract a **single primary**ⁱ when it is not possible to determine if there is a **single** melanoma or **multiple** melanomas.

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

Note 2: Examples of cases with minimal information include

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

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

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

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Single Melanoma

Note 1: Do not use the rules for melanoma described as metastasis

Note 2: Includes combinations of in situ and invasive

Rule M2 Abstract a **single primary**ⁱ when there is a **single melanoma**.

Note 1: A single melanoma is always a single primary

Note 2: The tumor may overlap onto or extend into adjacent/contiguous site or subsites.

Note 3: The tumor may have in situ and invasive components.

Note 4: The tumor may have two or more histologic components.

This is the end of instructions for Single Melanoma.

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

Multiple Melanomas

Note 1: Multiple melanomas may be a single primary or multiple primaries

Note 2: Do not use the rules for melanoma described as metastasis

Note 3: Includes combinations of in situ and invasive

Rule M3 Abstract **multiple primaries**ⁱⁱ when there are separate, non-contiguous melanomas in sites with ICD-O site codes that **differ** at the second (CXxx), third (CxXx) or fourth (C44X) character.

Note: This applies to a melanoma of unknown primary and a known cutaneous melanoma primary

Rule M4 Abstract **multiple primaries**ⁱⁱ when there are separate, non-contiguous melanomas with **different lateralities**.

Note 1: A **midline** melanoma is a different laterality than right or left.

Note 2: If the laterality of one or both melanomas is **unknown**, then continue through the rules

Note 3: If one or more of the sites is not in the **required laterality** table, then continue through the rules. See [Table 1](#).

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.

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

- Rule M5** Abstract **multiple primaries**ⁱⁱ when separate/non-contiguous tumors are two or more different subtypes/variants in Column 3, [Table 2](#) in the Equivalent Terms and Definitions. Timing is irrelevant.
Example: Epithelioid cell melanoma 8771/3 and nodular melanoma 8721/3 are both subtypes/variants of melanoma, NOS 8720/3.
- Rule M6** Abstract a **single primary**ⁱ when synchronous, separate/non-contiguous tumors are on **the same row in [Table 2](#)** in the Equivalent Terms and Definitions. Tumors must:
- Have the same site code **AND**
 - Have the same laterality
- Note:** The same row means the tumors are:
- The same histology (same four-digit ICD-O code) **OR**
 - One is the preferred term (column 1) and the other is a synonym for the preferred term (column 2) **OR**
 - A NOS (column 1/column 2) and the other is a subtype/variant of that NOS (column 3)
- Example 1:** Both lesions are located on the right side of the body and sites are chest C445 and back C445. Histology for the chest lesion is melanoma, NOS (8720/3) and the back lesion is nodular melanoma (8721/3). Abstract a single primary. Refer to [illustration](#).
- Example 2:** Both lesions are located on the left leg. One lesion is spindle cell melanoma (8772/3) located on the front of the left hip C447. The other lesion is spindle cell melanoma (8772/3) located on the front of the left thigh C447. Refer to [illustration](#).
- Rule M7** Abstract **multiple primaries**ⁱⁱ when melanomas are diagnosed more than 60 days apart.
Example: An **invasive** melanoma that occurs **more than 60 days after** an **in situ** melanoma is a multiple primary.
Note 1: The purpose of this rule is to ensure that the case is counted as an incident (invasive) case when incidence data are analyzed.
Note 2: Abstract as multiple primaries even if the medical record/physician states it is recurrence or progression of disease.

Cutaneous Melanoma Multiple Primary Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Rule M8 Abstract a **single primary**ⁱ when melanomas do not meet any of the above criteria.
Note 1: Use caution when applying this default rule. Please confirm that you have not overlooked an applicable rule.
Note 2: When an invasive melanoma follows an in situ melanoma **within 60 days**, abstract as a single primary.

This is the end of instructions for Multiple Melanomas

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

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

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Priority Order for Using Documents to Identify Histology

IMPORTANT NOTES

1. Code the histology diagnosed *prior* to **neoadjuvant treatment**.

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

Note 2: Neoadjuvant treatment is any tumor-related treatment given prior to surgical removal of the malignancy.

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

2. Code the histology assigned by the pathologist or 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.

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Note 2: The pathologist's diagnosis from the pathology report is always reliable, so the final diagnosis is the second priority.

Note 3: The CAP protocol is a checklist which:

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

2. Tissue/pathology from a **metastatic** site

Note 1: Code the behavior /3.

Note 2: The tissue from a **metastatic** site often shows **variations** from the primary tumor. When it is the only tissue available, it is **more accurate** than a scan.

3. **Scans: MRI, CT, PET. There is no priority** order because scans are not a reliable method for **identifying** specific **histology(ies)**.

4. Code the histology **documented** by the physician when none of the above are available. Use the documentation in the following priority order:

- A. Treatment plan
- B. Documentation from Tumor Board
- C. Documentation in the medical record that refers to original pathology, cytology, or scan(s)
- D. Physician's **reference to** type of cancer (**histology**) in the medical record

Note 1: Code the specific histology when documented.

Note 2: Code the histology to 8000 (cancer/malignant neoplasm, NOS) or as stated by the physician when nothing more specific is documented

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Coding Histology

Note 1: The priority is to code the most specific histology. **DO NOT USE BREAST HISTOLOGY CODING RULES FOR THIS SITE.**

Note 2: Only use this section for one or more histologies within a single tumor.

Note 3: Do not use this section in place of the Histology Rules.

1. Code the **most specific** histology or **subtype/variant**, regardless of whether it is described as:

- A. The majority or predominant part of tumor
- B. The minority of tumor
- C. A component

Example 1: Diagnosis for a single melanoma is melanoma, NOS 8720 with the majority or predominant part of tumor being nodular melanoma 8721. Code the subtype/variant: nodular melanoma 8721.

Example 2: Diagnosis for a single melanoma is melanoma, NOS 8720 with the minority of tumor being amelanotic melanoma 8730. Code the subtype/variant: amelanotic melanoma 8730.

Example 3: Diagnosis for a single tumor is melanoma, NOS 8720 with a component of malignant desmoplastic melanoma 8745. Code the subtype/variant: malignant desmoplastic melanoma 8745.

Note 1: The terms above (A, B, C) must describe a **melanoma** in order to code a histology described by those terms.

Example: When the diagnosis is melanoma with a nodular **melanoma** component, code nodular melanoma 8721.

Negative Example: When the diagnosis is simply melanoma with a nodular component, code melanoma, NOS 8720. Do not assume this is a nodular melanoma.

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

2. Code the histology described as **differentiation** or **features/features of ONLY** when there is a specific ICD-O code for the “NOS with ___ features” or “NOS with ___ differentiation”.

Note: Do not code differentiation or features when there is no specific ICD-O code.

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

3. Code the specific histology described by **ambiguous terminology** (list follows) **ONLY** when A or B is true:

A. The only diagnosis available is **one histology** term described by ambiguous terminology

- CoC and SEER require reporting of cases diagnosed only by ambiguous terminology
- Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available/documented

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

B. There is a **NOS histology and a more specific** (subtype/variant) described by ambiguous terminology

- Specific histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.) **OR**
- Patient is receiving treatment based on the specific histology described by ambiguous term

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

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

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

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

List of Ambiguous Terminology

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

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

- Architecture
- Foci; focus; focal
- Pattern

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Single Melanoma or Multiple Melanomas Abstracted as a Single Primary

- Rule H1** Code the histology when only **one histologic type** is identified.
Note 1: Use [Table 2](#) to code histology. New terms and synonyms are included in **Table 2** and coding errors may occur if the table is not used.
Note 2: When the histology is **not listed** in **Table 2**, use the ICD-O and all **updates**.
Note 3: Submit a question to [Ask a SEER Registrar](#) when the histology code is not found in Table 2, ICD-O, or all updates.
- Rule H2** Code the invasive histology when there are **invasive and in situ** components.
- Rule H3** Code the **histologic type** when the diagnosis is **regressing melanoma and a histologic type**.
Example: The lesion has evidence of regressing melanoma, nodular melanoma type. Code 8721/3 (Nodular melanoma).
- Rule H4** Code **8723/3** (Malignant melanoma, regressing) when the diagnosis is **regressing melanoma**.
Example 1: Malignant melanoma with regression. Code 8723 malignant melanoma, regressing.
Example 2: Malignant melanoma with features of regression. Code 8720/3 melanoma NOS.
- Rule H5** Code the **histologic type** when the diagnosis is **lentigo maligna melanoma and a histologic type**.
Example: The diagnosis is lentigo maligna melanoma with desmoplastic melanoma, right arm. Code desmoplastic melanoma, 8745/3.
- Rule H6** Code **8742/3** (Lentigo maligna melanoma) when the diagnosis is **lentigo maligna melanoma with no** other histologic types.
- Rule H7** Code the **subtype/variant** when there is a **NOS** and a **single subtype/variant** of that NOS.
• Melanoma NOS 8720 and a subtype/variant of melanoma
Note: Use [Table 2](#) in the Equivalent Terms and Definitions to determine NOS and subtype/variant.

Cutaneous Melanoma Histology Rules
C440-C449 with Histology 8720 – 8780 (Excludes melanoma of any other site)
Rules Apply to Cases Diagnosed 1/1/2021 forward

Rule H8 When two or more melanoma subtype/variants are present in a single tumor, submit a question to [Ask A SEER Registrar](#) for coding instructions.

Note 1: Two or more melanoma subtype/variants identified in a single tumor is rare.

Note 2: The WHO Classification of Skin Tumors 4th Ed does not include ICD-O codes for tumors with mixed melanoma subtype/variants

This is the end of instructions for Single Melanoma or Multiple Melanomas Abstracted as a Single Primary.

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

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

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

The following section is to be used for Other Sites cases diagnosed 2007-2021.

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

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

EQUIVALENT OR EQUAL TERMS

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

DEFINITIONS

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

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

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

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

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

Foci: Plural of focus.

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

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

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

Multiple primaries: More than one reportable case.

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

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

Recurrence: This term has two meanings:

1. The reappearance of disease that was thought to be cured or inactive (in remission). Recurrent cancer starts from cancer cells that were not removed or destroyed by the original therapy.
2. A new occurrence of cancer arising from cells that have nothing to do with the earlier (first) cancer. A new or another occurrence, incidence, episode, or report of the same disease (cancer) in a general sense – a new occurrence of cancer.

Single primary: One reportable case.

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

DETERMINING MULTIPLE PRIMARIES FOR SOLID MALIGNANT TUMORS

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

A. General Information

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

B. How to Use the Multiple Primary Rules

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

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

STOP

Histologic Type ICD-O-3

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

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

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

Information about the 2007 Histology Coding Rules

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

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

How to Use the Rules

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

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

STOP

Priority order for using Documents to Code Histology

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

1. Pathology report:
 - a. From the **most representative** tumor specimen examined
 - b. From the **final diagnosis**

Note 1: Use information from **addenda** and **comments** associated with the final diagnosis to code the histology.

Note 2: A **revised/amended diagnosis** replaces the original final diagnosis. Code the histology from the revised/amended diagnosis.

Note 3: The new rules **limit** the information **to the final diagnosis**. The old rules allowed coding from information in the microscopic description. You will only use information from the microscopic portion of the pathology report when instructed to do so in one of the site-specific rules.
2. Cytology report.
3. When you do not have either a pathology report or cytology report:
 - a. Documentation in the medical record that references pathology or cytology findings
 - b. From mention of type of cancer (histology) in the medical record

Ambiguous Terms Used to Code Histology

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

Ambiguous terms that are characteristic (used to code histology)

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

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

General Instructions Histology Coding Rules

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

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

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

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

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

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

Introduction

For cases diagnosed 1/1/2007 to 12/31/2021

IMPORTANT INFORMATION ON SITES COVERED IN THIS MODULE:

For cases diagnosed 1/1/2007 to 12/31/2017:

The Other Sites Rules cover rectosigmoid, rectum and all sites not included in the site-specific rules.

For cases diagnosed 1/1/2018 forward, the following sites are no longer included in the Other Sites Rules:

Rectosigmoid C199

Rectum C209

Peripheral Nerves and Autonomic Nervous System C470-C479

Beginning 1/1/2018, rectosigmoid (C199) and rectum (C209) primaries are included in the 2018 Colon Solid Tumor Rules

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

Equivalent or Equal Terms

- Acinar adenocarcinoma, adenocarcinoma (For prostate primaries only)
- Adenocarcinoma, glandular carcinoma
- And; with
Note: “And” and “with” are used as synonyms when **describing multiple histologies** within a **single tumor**.
- Tumor; mass; tumor mass; lesion; neoplasm
 - The terms tumor, mass, tumor mass, lesion, and neoplasm are **not** used in a standard manner in clinical diagnoses, scans, or consults. **Disregard** the terms unless there is a **physician’s statement** that the term is malignant/cancer
 - These terms are used **ONLY** to determine multiple primaries
 - **Do not** use these terms for casefinding or determining reportability

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

Table 1: Paired Organs and Sites with Laterality

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

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

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 2: Mixed and Combination Codes

This table is used to determine mixed and combination codes ONLY

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

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

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

Other Sites Equivalent Terms and Definitions
Excludes Head and Neck, Colon, Lung, Melanoma of Skin, Breast,
Kidney, Renal Pelvis, Ureter, Bladder, Brain, Lymphoma and Leukemia

Column 1: Required Histology	Column 2: Combined with Histology	Column 3: Combination Term	Column 4: Code
Gyn malignancies with two or more of the histologies in column 2	Clear cell Endometrioid Mucinous Papillary Serous Squamous Transitional (Brenner)	Mixed cell adenocarcinoma	8323
Papillary and Follicular		Papillary carcinoma, follicular variant	8340
Medullary	Follicular	Mixed medullary-follicular carcinoma	8346
Medullary	Papillary	Mixed medullary-papillary carcinoma	8347
Squamous carcinoma and Adenocarcinoma		Adenosquamous carcinoma	8560
Any combination of histologies in Column 2	Myxoid Round cell Pleomorphic	Mixed liposarcoma	8855
Embryonal rhabdomyosarcoma	Alveolar rhabdomyosarcoma	Mixed type rhabdomyosarcoma	8902
Teratoma	Embryonal carcinoma	Teratocarcinoma	9081
Teratoma and one or more of the histologies in Column 2	Seminoma Yolk sac tumor	Mixed germ cell tumor	9085
Choriocarcinoma	Teratoma Seminoma Embryonal	Choriocarcinoma combined with other germ cell elements	9101

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 3: Changes to Previous SEER Site Grouping Table

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

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

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

For cases diagnosed 1/1/2007 to 12/31/2021

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 (Cxxx) 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** (Cxxx) and are **in** any one of the following primary sites are multiple primaries. **
- Anus and anal canal (C21_)
 - Bones, joints, and articular cartilage (C40_ - C41_)
 - Peripheral nerves and autonomic nervous system (C47_) (Cases diagnosed 1/1/2007 to 12/31/2017 ONLY)
 - Connective subcutaneous and other soft tissues (C49_)
 - Skin (C44_)
- Rule M13** A **frank** in situ or malignant **adenocarcinoma** and an in situ or **malignant** tumor in a **polyp** are a single primary.*
- Rule M14** **Multiple** in situ and/or **malignant polyps** are a single primary. *
Note: Includes all combinations of adenomatous, tubular, villous, and tubulovillous adenomas or polyps.

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

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

Note 1: The purpose of this rule is to ensure that the case is counted as an incident (invasive) case when incidence data are analyzed.

Note 2: Abstract as multiple primaries even if the medical record/physician states it is recurrence or progression of disease.

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

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

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

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

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

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

**** Prepare two or more abstracts. Use the histology coding rules to assign the appropriate histology code to each case abstracted.**

This is the end of instructions for Multiple Tumors.

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

For cases diagnosed 1/1/2007 to 12/31/2021

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
- The final diagnosis is mucinous/colloid or signet ring cell adenocarcinoma in a polyp or
- There is documentation that the patient had a polypectomy

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

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

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

*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.

Jump to [Equivalent Terms and Definitions](#)

Jump to [Multiple Primary Rules](#)

Other Sites Solid Tumor Rules

For use with cases 2007-2021

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

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

Single Tumor: Invasive Only
(All parts are invasive)

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

Note 1: Priority for using documents to code the histology

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

Note 2: Code the specific histology when documented.

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

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

Note: Code the behavior /3.

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

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

Rule H11 Code the histology when only **one histologic type** is identified

Note 1: Do not code terms that do not appear in the histology description.

Example: Do not code squamous cell carcinoma non-keratinizing unless the words “non-keratinizing” actually appear in the diagnosis.

Note 2: If this is a papillary carcinoma of the thyroid, go to Rule H14.

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

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

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

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

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

Note: The specific histology may be identified as type, subtype, predominantly, with features of, major, or with ___ differentiation.

The terms architecture and pattern are subtypes only for in situ cancer.

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

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

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

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

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.

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

- 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.
