

Chapter 2:

Determining the Number of Primary Tumors

Now that you have learned how to determine if a case is reportable, you must next determine if the case is a single or a multiple primary. Use the following rules when abstracting cases diagnosed January 1, 2018, and later.

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.

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.

IMPORTANT:

For the complete *SEER Solid Tumor Rules* see: <https://seer.cancer.gov/tools/solidtumor/>

To determine multiple primaries for hematopoietic cancers such as lymphoma and leukemia, use the hematopoietic website at: <https://seer.cancer.gov/seertools/hemelymph/>

Definitions

Note: Use the terms and definitions in the following table for all reportable cases except lymphoma and leukemia primaries (ICD-O-3 histology codes 9590-9992).

Bilateral	Relating to the right and left sides of the body or of a body structure; bilaterality is not an automatic indication of single or multiple primaries; consult the site-specific instructions.
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.
De novo	In cancer, the first occurrence of cancer in the body.
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 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 below.)
Multiple primaries	More than one reportable case for the same patient.
NED	Acronym for "no evidence of disease"; disease free.
Non-contiguous	Not touching along the boundary; not being in actual contact.
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 above.)
Recurrence <i>This term has two meanings:</i>	<ol style="list-style-type: none"> 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 for a patient.
Unilateral	Relating to one side of the body or one side of a body structure.

2018 Solid Tumor Rules - What You Need to Know

(Excludes lymphoma and leukemia M9590 – M9992)

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

Eight site groups have been revised for 2018. The 2018 General Instructions apply only to the revised sites listed below:

1. Head & Neck
2. Colon (includes rectosigmoid and rectum for cases diagnosed 1/1/2018 forward)
3. Lung
4. Breast
5. Kidney
6. Urinary sites
7. Non-malignant CNS
8. Malignant CNS and Peripheral Nerves

The 2007 Multiple Primary & Histology rules and the 2007 General Instructions are to be used for cases diagnosed 1/1/2007 to 12/31/2019 for the following site groups:

- ***Cutaneous melanoma.*** Site rules will be revised for 2020 implementation to incorporate information from the new WHO 4th Edition Tumors of Skin.
- ***Other Site***
 - 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 2020 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.

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

How to Use 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

General Instructions - Multiple Primaries for Solid Tumors

1. Use the Solid Tumor Rules at: <https://seer.cancer.gov/tools/solidtumor/> to determine the number of reportable **primaries** and to code **histology**. Do **not** use these rules to determine case reportability, stage, or tumor grade.
2. The rules are effective for cases diagnosed **January 1, 2018**, and after. Do not use these rules to abstract cases diagnosed prior to January 1, 2018.

Note: For tumors diagnosed 01/01/2007 through 12/31/2017, use the 2007 Multiple Primary and Histology (MP/H) Rules: <https://seer.cancer.gov/tools/mphrules>.
3. Read the **General Instructions** and the **site-specific Equivalent Terms and Definitions** in the Solid Tumor Rule manual before using the rules.
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 module. Use the first rule that applies and **STOP**.
6. **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 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.
7. These rules do not apply to tumors described as metastases.

IMPORTANT: Do **not** use Solid Tumor Rules to determine case reportability, casefinding, stage, or tumor grade.

General Instructions - Multiple Primaries Rules for Solid Tumors

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.

Note: When there is a tumor or tumors with separate microscopic foci, ignore the microscopic foci.
 - c. When the patient has a **single tumor**, use the “Single Tumor” module.
 - d. When the patient has **multiple tumors**, use the “Multiple Tumors” module.
2. When the rules return a single primary, prepare one abstract.
3. When the rules return multiple primaries, prepare two or more abstracts.
4. For those sites/histologies which have recognized **biomarkers**, the biomarkers frequently identify the histologic type. Currently, there are clinical trials being conducted to determine whether these biomarkers can be used to identify multiple primaries. Follow the Multiple Primary Rules; do not code multiple primaries based on biomarkers.

Timing Rules for Solid Tumors

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

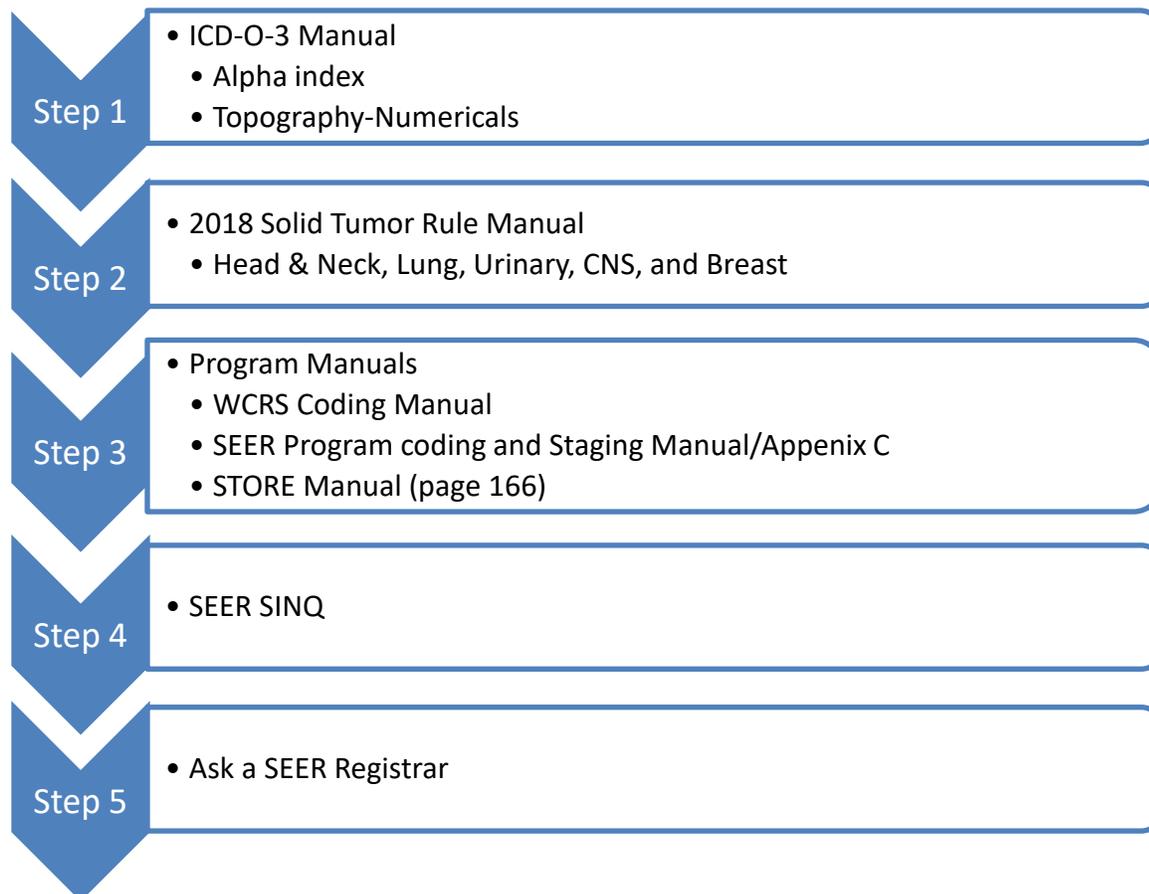
Examples of timing rules:

- Abstract **multiple primaries** when the patient has a subsequent tumor for the same site after being **clinically disease-free** for **greater than** the time frame interval as listed in the Solid Tumor Rule site-specific chapters (for example, the colon cancer interval is three years, where the invasive breast cancer interval is five years).

Note: If there is a recurrence less than or equal to the time frame interval as listed in the Solid Tumor Rule site-specific chapter, the **“clock”** starts over. The time interval is now calculated from this **date of last recurrence**, instead of the original date of diagnosis.

- Abstract a **single primary** (the invasive) when an **invasive** tumor is diagnosed **less than or equal to 60 days after** an **in-situ** tumor
- **Clinically disease-free** means that there was **no evidence** of recurrence on follow-up.
- 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”.

Steps for Coding Primary Site for Solid Tumors



Step 1

<http://codes.iarc.fr/>

Step 2

https://seer.cancer.gov/tools/solidtumor/STM_2018.pdf

Step 3

<https://www.dhs.wisconsin.gov/wcrs/reporterinfo/manual.htm>

<https://seer.cancer.gov/tools/codingmanuals/index.html>

https://www.facs.org/~media/files/quality%20programs/cancer/ncdb/store_manual_2018.ashx

Step 4

<https://seer.cancer.gov/seerinqury/index.php>

Step 5

<https://seer.cancer.gov/registrars/contact.html>

General Information - Coding Histologic Type for Solid Tumors

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

The update includes:

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

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

- Updated ICD-O histology codes and terms: <https://seer.cancer.gov/icd-o-3/>
- The 2018 *Solid Tumor Rules*
- The ICD-O

When a histology code cannot be identified using the above recommendations, review *SEER SINQ* to see if this scenario has been previously submitted and answered by SEER. If not, submit a question to “*Ask a SEER Registrar.*”

General Instructions - Histology Coding Rules for Solid Tumors

Note 1: Do not use these rules to determine case reportability.

Note 2: Refer to *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.

Note 1: Each section is a complete set of rules.

Note 2: 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 to **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:
 - a. Histology is clinically confirmed by a physician (attending, pathologist, oncologist, etc.)
 - b. Patient is treated for the histology described by an ambiguous term.
 - c. Case is accessioned (added to your database) based on ambiguous terminology and no other histology information is available or documented.

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

Ambiguous Terminology for Solid Tumor Histology

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

IMPORTANT: Ambiguous terminology to determine histology for Solid Tumor Rules is NOT the same as ambiguous terminology used to determine reportability.

Priority Order for Using Documents to Code Histology

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

- Priority order will differ by site. Biomarkers and/or tissue pathology always takes precedence.
- The specific types of radiography/scans also differ by site.

General Information – Coding Hematopoietic and Lymphoid Neoplasm Multiple Primary Tumors and Histology

The Hematopoietic & Lymphoid Neoplasm Database (Heme DB) contains abstracting and coding information for all hematopoietic and lymphoid neoplasms (9590/3-9992/3). The Coding Manual contains reportability instructions and rules for determining the number of primaries, the primary site and histology.

IMPORTANT: For lymphomas, leukemia and other hematopoietic malignancies, primary site and timing are not applicable for determining single or multiple primaries – histology becomes the determining factor.

Refer to the Heme DB and Coding Manual for complete instructions on the SEER website at:

<http://seer.cancer.gov/seertools/hemelymph/>

Steps in Priority Order for Using the Hematopoietic and Lymphoid Neoplasm Database and Coding Manual

1. Identify the working (preliminary) **histology code(s)** by searching the Heme DB.

Examples of Searches		
Search	Example	Notes
Unique word in diagnosis	“precursor”	If diagnosis is precursor acute lymphoblastic leukemia
Complete name (diagnosis)	“acute myelomonocytic leukemia”	The number of matched terms will be much smaller than just searching on “leukemia”. Results displayed will have all three words in the histology name. The words may appear in any part of the entry (alternative names, abstractor notes, transformations, etc.
Abbreviations	AMML	Acute myelomonocytic leukemia
Histology code	9867/3	Code search display’s: Acute myelomonocytic leukemia

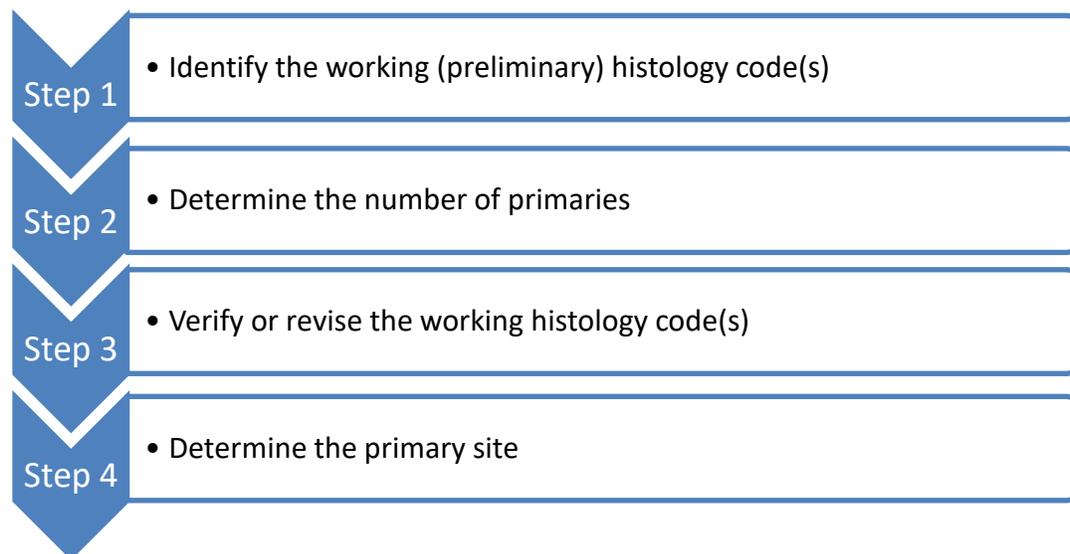
Note: When multiple results are displayed, click on the desired term to display the record.

2. Use the Multiple Primary Rules to determine the **number of primaries** using the working histology code(s).
 - a. Start with rule M1, move through the rules in consecutive order and stop at the first rule that applies. The M rule references in the Heme DB are to be used as a guide only.
 - b. Use the Hematopoietic Multiple Primaries Calculator in the Heme DB only when instructed by the rules in the Hematopoietic Manual.
3. Verify or revise the working histology code(s) using the Primary Site and Histology Rules.
 - a. When the PH rules lead you to a different histology code, enter that code in the Heme DB search box and display the record for that histology.
 - b. The PH rules referenced in the Heme DB are the most common rule(s) used to code Primary Site and Histology for the selected histology. More than one Module/PH Rule may be needed to code Primary Site and Histology.
4. Determine primary site using the *Primary Site and Histology Rules*
 - a. See Primary Site Coding Instructions.
 - b. For certain histologies, only one primary site code is displayed.
 - c. When there is no primary site code listed under **Primary Site(s)**
 - Review the **Primary Site Text** field for common primary sites or other primary site instructions and rules.
 - Search the Hematopoietic Manual and/or database to find applicable modules.
 - Read the **Abstractor Notes** to find other information regarding sites of involvement for stages II, III, and IV lymphomas. Use the **Abstractor Notes** to confirm that the **site/histology combination indicated by the involvement documented in the medical record is probable**.
 - You may also seek a physician's help in determining the primary site.

IMPORTANT: Grade is no longer applicable for Hematopoietic and Lymphoid Neoplasms for cases diagnosed 2018 and forward.

- **For cases with histologies 9590/3-9992/3, the clinical grade must be coded to '8'.**
- **Grade fields are coded to '8', not applicable**
 - **Exception:** Follicular lymphomas occurring in the Lymphoma Ocular Adnexa schema.

Steps for Using the Hematopoietic and Lymphoid Neoplasm Database



Example: A patient is diagnosed at your facility in 2018 with acute myeloid leukemia. The patient is in your registry database with refractory anemia with ring sideroblasts diagnosed and treated in 2010.

Step 1: Identify the working histology code (s).

- Refractory anemia with ring sideroblasts – 9982/3
- Acute myeloid leukemia – 9861/3

Step 2: Determine the number of primaries.

- Rule M10: Abstract as multiple primaries when a neoplasm is originally diagnosed as a chronic neoplasm AND there is a second diagnosis of an acute neoplasm more than 21 days after the chronic diagnosis.

Note: Check the Hematopoietic Database to determine if histology is transformation to/from.

Step 3: Verify or revise the working histology code (s).

- 2010 – 9982/3
- 2018 – 9861/3

Step 4: Determine the primary site.

- 2010 - C421
- 2018 – C421