2010 Hematopoietic and Lymphoid Neoplasm
Case Reportability and Coding Manual
Effective with Cases Diagnosed 1/1/2010 and after

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This manual and the companion Hematopoietic and Lymphoid Neoplasm Database (Hematopoietic DB) are dedicated to the Hematopoietic Working Group whose tireless efforts have made this project a success. We also dedicate these new cancer reporting tools to the hard-working cancer registrars across the world who meticulously identify, abstract and code cancer data; and are the foundation for statewide, provincial, territorial, national, and international cancer surveillance programs which support cancer prevention and cancer control efforts worldwide. We also wish to recognize the assistance, support and significant contributions of the Statistical Review Working Group.

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Introduction

Background

The Hematopoietic Working Group was led by the Surveillance, Epidemiology, and End Results (SEER) Program of the National Cancer Institute (NCI) and included members from many professional organizations: the National Cancer Registrars Association (NCRA), the North American Association of Central Cancer Registries (NAACCR), the National Program of Cancer Registries (NPCR) of the Centers for Disease Control and Prevention (CDC), the Commission on Cancer (CoC) of the American College of Surgeons (ACoS), and the Canadian Cancer Registries (CCR). The Working Group also included cancer registrars who work independently (contractors), hospital registrars, central cancer registry registrars, and clinical and research physicians who are experts in the hematopoietic and lymphoid neoplasm fields.

This working group has developed rules, guidelines and an interactive desktop Hematopoietic and Lymphoid Database (Hematopoietic DB) reference to assist registrars in determining case reportability, the number of primaries, and how to code primary site, histology, and grade for a hematopoietic and/or lymphoid neoplasm. The rules, guidelines, and the Hematopoietic DB follow the *World Health Organization (WHO) Classification of Tumours of the Haematopoietic and Lymphoid Tissues*, 4th Edition, 2008, also called the "WHO Blue Book." Both the *International Classification of Diseases for Oncology (ICD-O)* and the series of Blue Books are produced by the WHO, but the content of the books are very different. Each has a prominent place in the registry world.

The original ICD-O, the ICD-O-2, and the ICD-O-3 provide standard primary site and histology codes for specific benign, borderline, and malignant conditions. The ICD-O series also provides generic "not otherwise specified" or "NOS" codes for some conditions so registrars are able to code cases that have limited information, such as death-certificate-only cases, historic cases, and other cases for which specific information is not available. When ICD-O attributes a code to a specific histology, it is a very rare occasion for that original code to be changed. The intent is that the code should never change; for example code 8140/3 for adenocarcinoma, NOS has remained unchanged since the first edition of ICD-O. These manuals are the standard for coding neoplasms throughout the world. To preserve the integrity of historical data, and to allow for comparison of data over time, it is imperative that standard codes remain unchanged. Although the stability of these codes is necessary to interpret data over time, it has some less-than-desirable results. When the clinical world reclassifies diseases to reflect the current state of science and knowledge about a particular disease or condition, that disease will remain in the same numerically controlled category (rubric) in ICD-O. When the ICD-O editors assign new codes for a neoplasm, the new code may not be placed in the proper category because unassigned codes under the correct histology grouping may not be available. An example of this problem is the placement of the non-Hodgkin lymphomas that were first added in ICD-O-3.

The WHO Blue Books, by contrast, are histo-pathology reference books used by pathologists and oncologists throughout the world. The Blue Books are revised and published when new information is available. The *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition, a collaborative project of the Society for Hematopathology/European Association for Haematopathology was published in 2008. The reference includes new disease classifications, changes to existing classifications and cell lineages, and new conditions that reflect the state-of-the-science for these diseases. This reference was the primary source of information used to develop the *2010 Hematopoietic and Lymphoid Neoplasm Case Reportablility and Coding Manual* and the accompanying Hematopoietic DB because the WHO Blue Book is consistently updated with the current classification by cell lines or lineages and classification groupings. Using the WHO classifications gives the registrar reference material that is clinically relevant and compatible with the information being abstracted from the medical records. When the clinical field finds specific tumor markers, immunohistochemical testing, genetic testing, or other characteristics that define or refine a diagnosis or a particular histology, the WHO Blue Books introduce proposed new codes for new or more specific histologies, and these new histologies may be grouped or classified under a different or a new category based on information about the origins or behavior of the neoplasm. It takes time for these

proposed new terms and codes to be published in an addendum to the ICD-O or in a revision of the ICD-O. Thus, it is not unusual for the registrar to see a pathologist use the proposed new terminology or condition code as presented in the Blue Book. Prior to 2010 registrars were instructed to use only those codes listed in the most current version of the ICD-O.

The *Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual* and Hematopoietic DB are designed to help the registrar understand and interpret the information written by pathologists. The Hematopoietic DB will be updated periodically to ensure that the registrar has the most current information available to interpret and code a hematopoietic or lymphoid neoplasm.

The Hematopoietic Working Group requested a review and approval for implementing the new terms and corresponding ICD-O codes as well as the newly reportable neoplasms from the Cancer Registration Steering Committee (CRSC). After review and discussion the CRSC approved the use of the new codes and terms and also the collection of the newly reportable neoplasms for cases diagnosed 1/1/2010 and after. If needed, it is recommended that central registries revise their legislation to include these newly reportable neoplasms.

***Note**: The WHO is the directing and coordinating authority for health within the United Nations system. It is responsible for providing leadership on global health matters, shaping the health research agenda, setting norms and standards, articulating evidence-based policy options, providing technical support to countries and monitoring and assessing health trends. (From http://www.who.int/about/en/)*

Diagnostic Confirmation
Code bone marrow aspiration, bone marrow biopsy, CBC, and peripheral blood smear as positive histologic confirmation (code 1).

Do **not** use the instructions that direct you to give preference to coding a histologic confirmation for the hematopoietic and lymphoid neoplasms. For these neoplasms, the genetic testing and immunophenotyping are needed to identify the specific histology. When the Hematopoietic DB lists genetic testing, immunophenotyping, or any other variation of laboratory testing as the definitive diagnostic method **and** any one of those tests were done to confirm the histologic type, code diagnostic confirmation as positive laboratory test/marker study (code 5).

Diagnostic Process for Leukemia
For most patients, the first suspicion or presentation of a hematopoietic or lymphoid neoplasm is the clinical scenario, including symptoms such as unexplained weight loss, weakness, chronic fatigue, easy bruising, etc. For leukemia, the physician usually orders a complete blood count (CBC) and/or a peripheral blood smear because these tests will identify whether the abnormalities occur in the white blood cells or the red blood cells. Secondly a bone marrow (BM) biopsy is usually conducted so the bone marrow can be studied under the microscope. There are a few neoplasms where the CBC or peripheral blood count alone or the bone marrow biopsy provides a definitive diagnosis. However, in most cases the results of these tests will provide several potential diagnoses which may include one or more of the non-specific or NOS hematopoietic lymphoid neoplasm categories: myeloproliferative neoplasms, myeloid and lymphoid neoplasms, myelodysplastic/myeloproliferative neoplasms, myelodysplastic syndromes, acute myeloid leukemia, acute leukemia of ambiguous lineage, precursor lymphoid neoplasms, mature B-cell neoplasms, mature T-cell and NK-cell neoplasms, histiocytic and dendritic cell neoplasms, and post-transplant lymphoproliferative disorders (Appendix B). Appendix B can be used as a reference to identify which diseases or conditions are included in a specific classification group or category. The physician may suggest several potential specific diagnoses (differential or provisional), indicating that more testing is needed to identify the specific hematopoietic or lymphoid neoplasm and subsequent treatment. The new WHO classification of diseases defined in the 4th Edition requires more extensive immunophenotyping and, in some cases, genetic information for classification. The Hematopoietic DB contains specific information on the types of diagnostic tests that are used to identify the specific histology for the hematopoietic or lymphoid neoplasm being abstracted. See “Definitive Diagnostic Method” in the Hematopoietic DB.

Leukemia vs. Lymphoma
The difference between leukemia and lymphoma is that leukemia most commonly presents in the bone marrow and/or blood while lymphoma most commonly manifests in lymph nodes, lymphoid tissue or lymphoid organs. When only the bone marrow is involved, the histology is usually leukemia; rarely a lymphoma may present only in the bone marrow (See the PH rules for instructions on coding primary site for lymphomas). Both leukemia and lymphoma patients may have splenomegaly (enlargement of the spleen). Patients with leukemia may have leukemic infiltrate of the spleen. Splenomegaly does not mean that the leukemia originated in the spleen or that this neoplasm is lymphoma.

Transformation
Although the process of transformation from a chronic to an acute leukemia is appropriately called a progression of disease, it is not comparable to the progression of disease in solid tumors. Solid tumors progress by extension, seeding, or blood-born micrometastases to regional and/or distant sites. The solid tumor disease progression is the same histology as the original tumor; metastatic sites that are biopsied show the same histology as the original tumor. In contrast, when the chronic phase of leukemia progresses or transforms to the acute phase, the histology actually changes. The acute phase is a much more aggressive manifestation of the disease process. Treatment and survival for chronic and acute leukemia is vastly different. (See Multiple Primary Rules for information on how to code transformations.)

Role of the Hematopoietic Database (Hematopoietic DB)
The new Hematopoietic DB will enable registrars to identify and understand a hematopoietic or lymphoid neoplasm as well as correctly and consistently abstract and code cases. Users will be able to query any final, differential, or provisional diagnosis in the Hematopoietic DB. Once the condition has been identified, the diagnostic or confirmatory tests will be listed under "definitive diagnosis" for each neoplasm. This will provide the information needed to search the medical record for specific diagnostic test results. Some healthcare institutions may "file" confirmatory test results such as immunophenotyping or genetic testing in a location other than the location used for standard laboratory tests in the medical record. We recommend that the registrar ask the laboratory for examples of printed test results such as immunophenotyping or genetic testing to become familiar with the format of the test results as well as other information that may be included with test results. We also recommend that the registrar ask the Health Information Management or Medical Records Department where these tests are "filed" within the chart (paper or electronic).

Information for Lymphoma Only
The anatomic site that is most accessible or easiest to reach is usually biopsied when a patient has abnormal lymph nodes and a diagnostic biopsy is warranted. For example, if a CT or PET scan identified enlarged cervical and mediastinal lymph nodes, the physician would biopsy the cervical lymph nodes. A mediastinal lymph node biopsy would require puncturing the chest cavity while cervical lymph nodes can be biopsied with a local anesthetic. The less invasive procedure is used to confirm the diagnosis and minimize the risk of complication, infection, and trauma to the patient. This does not imply that the disease process started in the cervical lymph nodes.

Hodgkin lymphoma (HL) is a type of lymphoma originating in lymphocytes (a type of white blood cell). HL is characterized by the presence of Reed-Sternberg cells (RS cells) on microscopic examination. HL usually originates in the lymph nodes and is characterized by the orderly spread of disease from one lymph node group to another. The patient develops systemic symptoms with advanced disease (metastasis) to the spleen, liver and/or bone marrow.

Non-Hodgkin lymphoma (NHL) comprises a diverse group of malignant neoplasms which include all lymphomas other than Hodgkin. NHL arises in lymphocytes (white blood cells). Lymphocytes are present in lymph nodes and throughout the body. NHL commonly develops in lymph nodes but also occurs in extranodal sites including: tonsils, spleen, ileum, stomach, Waldeyer ring, bone marrow, skin, bone, central nervous system, lung, gonads, conjunctiva, ocular adnexae, liver, kidneys, and uterus.

What is New and/or Different in the 2010 Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Rules?

- *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition is the authoritative reference used to develop the rules, the information in the Hematopoietic DB, the preferred histologic term, the histologic groupings, the new histology codes and terms, and the newly reportable diseases. The *International Classification of Diseases for Oncology*, 3rd Edition (ICD-O-3) was the authoritative reference for the remainder of the histology terms and for the synonyms.
- Changes in Reportability
    - New ICD-O histology terms and codes from *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition (33 codes/histologies)
    - Changes to existing codes from non-reportable /1 to reportable /3 (3 codes/histologies).
    - Transformations collected as new primaries.
- For cases diagnosed 2010 and later, the *Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual* and the Hematopoietic DB replace:
    - The February 2001 *Single Versus Subsequent Primaries of Lymphatic and Hematopoietic Diseases* table
    - *Abstracting and Coding Guide for the Hematopoietic Diseases*, 2002.
    - Previous casefinding and Reportable Neoplasm lists for hematopoietic neoplasms (ICD-9-CM and ICD-10).
- The *Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual* provides:
    - Case Reportability Instructions that include guidance on descriptive phrases and newly reportable conditions.
    - Multiple Primary Rules that reinforce the instructions to abstract the acute and chronic phases of the same disease as separate primaries.
    - Primary Site and Histology Coding Rules that guide the registrar in coding the correct primary site and the most appropriate and most specific histology rather than the histology at diagnosis.
    - Grade Coding Rules that guide the registrar to code the cell line origins including coding the specific type when both null cell and a specific type such as T-cell are stated in the diagnosis.

Case Reportability Instructions

Note 1: In many cases the registrar will need to make inquiries to the physician's office to confirm the diagnosis. Unless that type of follow-back is done, hematopoietic cases will be under-reported.

Note 2: When a **pathology report provides the final diagnosis**, report the most specific histology recorded in any of the following parts of the pathology report

- As the final diagnosis
- In a comment regarding the final diagnosis
- As an addendum to the final diagnosis
- In the College of American Pathologists (CAP) protocol

Note 3: Reportable diagnoses are listed in Case Reportability Instructions 4-10

1. Report the case when the only information available is that the clinician has started **cancer-directed treatment** for a reportable hematopoietic or lymphoid neoplasm described in Case Reportability Instructions 4-10
   Note 1: Report the case even if the diagnostic tests are inconclusive, equivocal, or negative.
   Note 2: For cancer-directed treatment information see the National Cancer Institute's Physicians' Data Query (PDQ) website at http://www.nci.nih.gov/cancertopics/pdq/cancerdatabase

2. Report the case when the diagnosis of a hematopoietic or lymphoid neoplasm is preceded by one of the following **ambiguous terms**
   Note: Do **not** report cases diagnosed only by ambiguous **cytology** (cytology diagnosis preceded by ambiguous term).
   - Apparent(ly)
   - Appears
   - Comparable with
   - Compatible with
   - Consistent with
   - Favor(s)
   - Malignant appearing
   - Most likely
   - Presumed
   - Probable
   - Suspect(ed)
   - Suspicious (for)
   - Typical (of)

   Note 1: Reportable diagnoses are described in Case Reportability Instructions 4-10
   Note 2: Use these terms when screening all diagnoses other than cytology and tumor markers.
   Note 3: Report only those cases that use the words on the list or an equivalent word such as "favored" rather than "favor(s)". Do not substitute synonyms such as "supposed" for "presumed" or "equal" for "comparable."
   Note 4: Accept the reportable term and report the case when one part of the medical record uses a reportable ambiguous term such as "apparently" and another section of the medical record(s) uses a term that is not on the reportable list.

Note 5: Diagnoses based on ambiguous terminology require follow-back to see if the diagnosis has been confirmed or proven to be incorrect (see note 6).
Note 6: Do **not** report the case when biopsy or physician's statement proves the ambiguous diagnosis is **wrong** (for example, pathology diagnosis is benign or borderline).
Example: CT scan shows enlarged lymph nodes suspicious for lymphoma. Subsequent biopsies of the lymph nodes thought to be involved with a neoplasm are negative for malignancy. The pathology is more reliable than the scan; the negative biopsy proves that the presumed malignancy does not exist. Do **not** report the case.

3. Report the case when there is a **clinical diagnosis** (physician's statement) of reportable hematopoietic or lymphoid neoplasm.
   Note 1: Reportable diagnoses are listed in Case Reportability Instructions 4-10.
   Note 2: The clinical diagnosis may be a final diagnosis, found within the medical record or recorded on a scan (CT, MRI for example).
   Note 3: Report the case even if the diagnostic tests are equivocal. A number of hematopoietic diseases are "diagnoses of exclusion" in which the diagnostic tests are equivocal and the physician makes the clinical diagnosis based on the equivocal tests and the clinical picture. See the Hematopoietic DB for definitive diagnostic procedures for the specific disease being abstracted.

4. Report the case when **multiple myeloma, evolving myeloma, early multiple myeloma, indolent multiple myeloma** or **smoldering multiple myeloma** is diagnosed.

5. Report the case when **preleukemia** or **smoldering leukemia** is diagnosed.
   Note: In ICD-O-3 preleukemia is listed as 9989/3 in the numeric list and 9989/1 in the alphabetic list/index. Change the 9989/1 in the alphabetic list to a 9989/3 in your ICD-O-3.

6. **Report** the following hematopoietic and lymphoid neoplasms **as malignant**
   - Langerhans cell histiocytosis, NOS (9751/3)
   - Myeloproliferative neoplasm, unclassifiable / myelodysplastic/myeloproliferative neoplasm unclassifiable (9975/3)
   - T-cell large granular lymphocytic leukemia/chronic lymphoproliferative disorder of NK cells (9831/3)

   Note: This is a change from previous rules. These neoplasms are listed in ICD-O-3 as uncertain whether benign or malignant /1 but were changed to reportable /3 in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition. (See Appendix D for more information).

7. Report the case when a reportable diagnosis appears in any text or report described as a **definitive diagnostic method** in the Hematopoietic DB.
   Note 1: Reportable diagnoses are listed in Case Reportability Instructions 4-10.
   Note 2: Definitive diagnostic methods differ depending upon the histology. See the Hematopoietic DB for details.

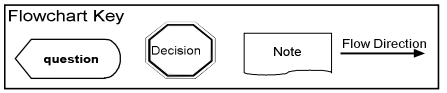
8. Report hematopoietic and lymphoid neoplasms with ICD-O-3 morphology codes **9590-9992** that are listed as **/1** and **described as malignant** by a physician.
   Note: There are **no** in situ (/2) hematopoietic or lymphoid neoplasms.

9. Report all ICD-O-3 morphology codes **9590-9992** with a **/3** behavior plus the **new histology terms and codes** published by *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition (See Appendix D for complete list).
   Note: These terms are not listed in the ICD-O-3 implemented in 2001. The new WHO codes allow these neoplasms to be coded as a specific disease rather than one of the NOS categories. Use the codes in Appendix D until ICD-O-4 is published or an addendum to ICD-O-3 is distributed.

10. Query the **Hematopoietic DB** to determine case reportability for special cases that do **not** meet the criteria listed in the above instructions.

Multiple Primary Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use Multiple Primary Rules M1 through M12 before using the Hematopoietic DB.



*Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.
**Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

UNKNOWN IF SINGLE OR MULTIPLE TUMORS	DECISION	NOTES and EXAMPLES
		The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules M7 - M12.
M1 Is there **minimal information** available (such as a DCO case or a pathology-report-only case).	YES → SINGLE Primary*	
NO		
M2 Is there a **single histology**?	YES → SINGLE Primary*	***Example 1:*** The diagnosis is multiple myeloma (9732/3). Abstract as a single primary. ***Example 2:*** Multiple extraosseous plasmacytomas (9734/3) are present in the oropharynx. Abstract as a single primary. ***Example 3:*** A single histology diagnosed by the definitive diagnostic method as defined in the Hematopoietic DB; **for example.** The patient had several provisional diagnoses but the definitive diagnostic method identifies a single histology. Abstract as a single primary.
NO		
M3 Are there **two or more types of non-Hodgkin lymphoma** present in the **same anatomic location(s),** such as one lymph node, one organ, or one tissue?	YES → SINGLE Primary*	***Example 1:*** Biopsy of cervical lymph node shows **two** different non-Hodgkin lymphomas. Abstract as a single primary. 1. Do NOT use this rule for cutaneous lymphomas. 2. When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for information on lymph node codes and regions. 3. When the disease is in a more advanced stage, both non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3or in an organ and that organ's regional lymph nodes or in multiple organs. ● Although both non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination non-Hodgkin lymphoma, assume that all of the nodes, tissue, and/or organs are involved with the combination of non-Hodgkin lymphomas. 4. Do not query the Hematopoietic DB in this situation.
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Multiple Primary Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use Multiple Primary Rules M1 through M12 before using the Hematopoietic DB.

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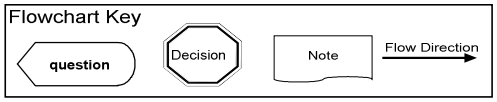
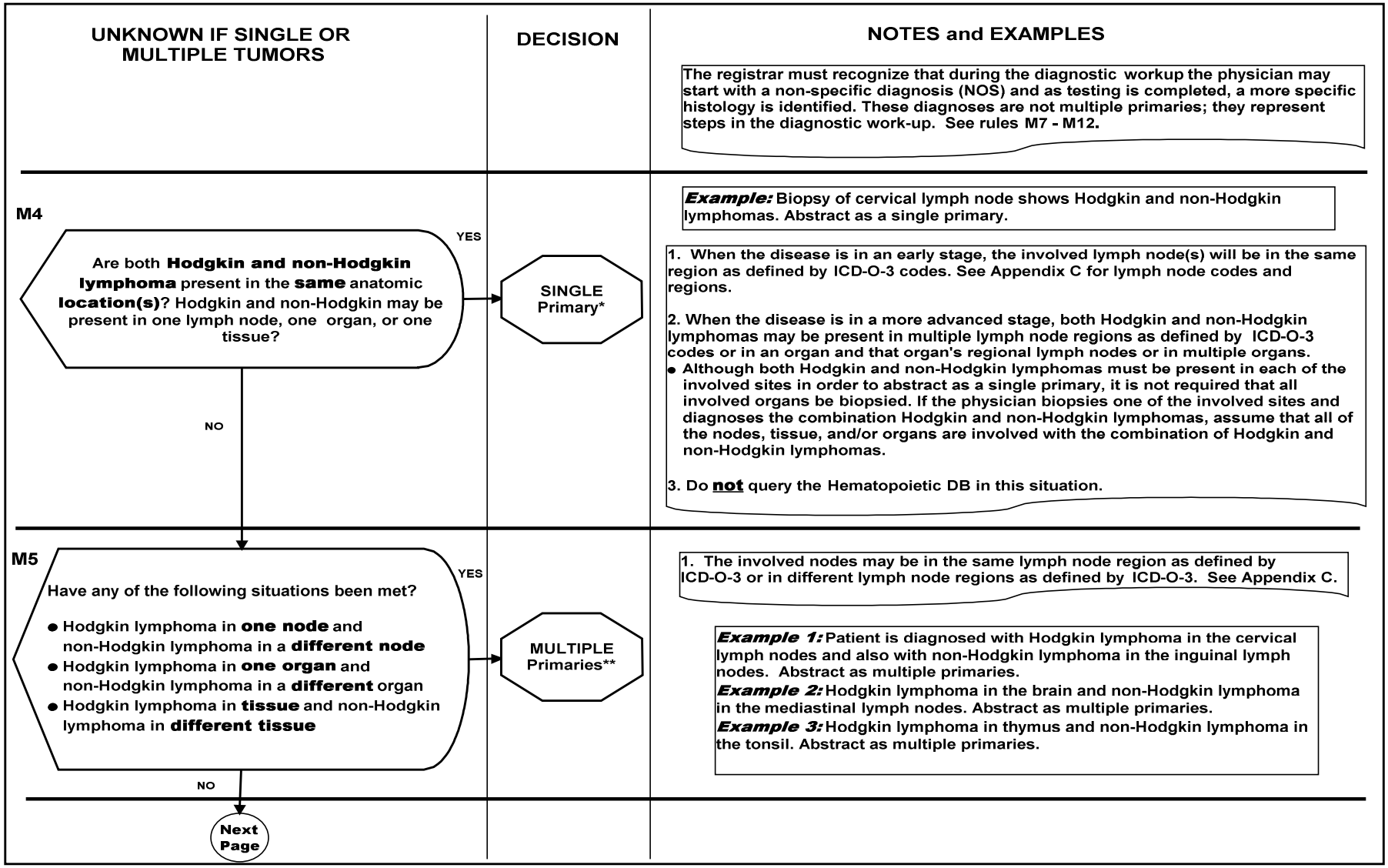
**Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.





Multiple Primary Rules - Flowchart
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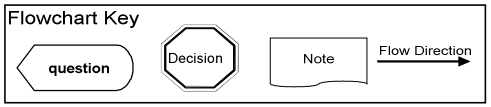
Use Multiple Primary Rules M1 through M12 before using the Hematopoietic DB.



***Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.**
****Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.**

UNKNOWN IF SINGLE OR MULTIPLE TUMORS	DECISION	NOTES and EXAMPLES
		The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules M7 - M12.
M6 Is there more specific histology diagnosed after an NOS when the Hematopoietic DB Multiple Primary Calculator confirms that the NOS and more specific histology are the same primary? YES → NO	SINGLE Primaries*	1. There are no time restrictions on these diagnoses; the interval between the NOS and the more specific histology does not affect this rule stating that the two neoplasms are a single primary. 2. The Hematopoietic DB will identify these histologies as a single primary.
M7 Are both the chronic and the acute phase of the neoplasm diagnosed within 21 days **AND** there is documentation of one positive bone marrow biopsy? YES → NO	SINGLE Primaries*	When these diagnoses happen within 21 days, it is highly possible that one diagnosis was provisional and the bone marrow identified the correct diagnosis.
M8 Are both the chronic and the acute phase of the neoplasm diagnosed within 21 days **AND** there is documentation of two bone marrow examinations, one confirming the chronic neoplasm and another confirming the acute neoplasm? YES →	MULTIPLE Primaries**	
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Multiple Primary Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

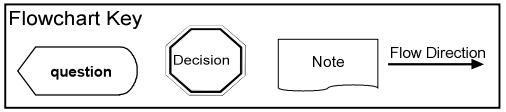
Use Multiple Primary Rules M1 through M12 before using the Hematopoietic DB.

*Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.
**Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

UNKNOWN IF SINGLE OR MULTIPLE TUMORS	DECISION	NOTES and EXAMPLES
		The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules M7 - M12.
M9 Are both the chronic and the acute phase of the neoplasm diagnosed within 21 days **AND** there is no available documentation on bone marrow biopsy?	YES → SINGLE Primary*	1. The two diagnoses are likely the result of an ongoing diagnostic work-up. The later diagnosis is usually based on all of the test results. 2. This rule applies if both neoplasms are diagnosed simultaneously (at the same time).
M10 Is a neoplasm **originally diagnosed** in a **chronic** (less aggressive) phase **AND second diagnosis** of a blast or acute phase **21 days or more** after the chronic diagnosis?	YES → MULTIPLE Primaries**	1. **This is a change from previous rules.** Use the Hematopoietic DB to determine multiple primaries when a transformation from the chronic to a blast or acute phase occurs. 2. Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.
NO → Next Page		

Multiple Primary Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use Multiple Primary Rules M1 through M12 before using the Hematopoietic DB.



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UNKNOWN IF SINGLE OR MULTIPLE TUMORS	DECISION	NOTES and EXAMPLES
		The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules M7 - M12.
M11 Is a neoplasm **originally diagnosed** in the blast or acute phase and **reverts** to a less aggressive/chronic phase and there is **no confirmation** available that the patient has been treated? YES	SINGLE Primary*	1. When these diagnoses happen **within 21 days**, it is highly possible that the first diagnosis of acute disease was a provisional diagnosis. 2. When the subsequent diagnosis occurs more than 21 days after the original diagnosis of acute disease it is important to follow-back to obtain information on treatment or a subsequent bone marrow biopsy that negates the diagnosis of acute disease.
NO		
M12 Is a neoplasm **originally diagnosed** in the blast or acute phase and **reverts** to a less aggressive/chronic phase **after treatment**? YES	MULTIPLE Primaries**	1. Only abstract as a multiple primary when the patient has been treated for the acute disease. 2. **This is a change from previous rules.** Use the Hematopoietic DB to determine multiple primaries when a transformation from the blast or acute phase to a chronic phase occurs. 3. Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.
NO		
M13 Is it impossible to determine the number of primaries for all cases using criteria M1-M12? YES	Use the **Hematopoietic DB**	

This is the end of instructions for the Multiple Primary Rules.

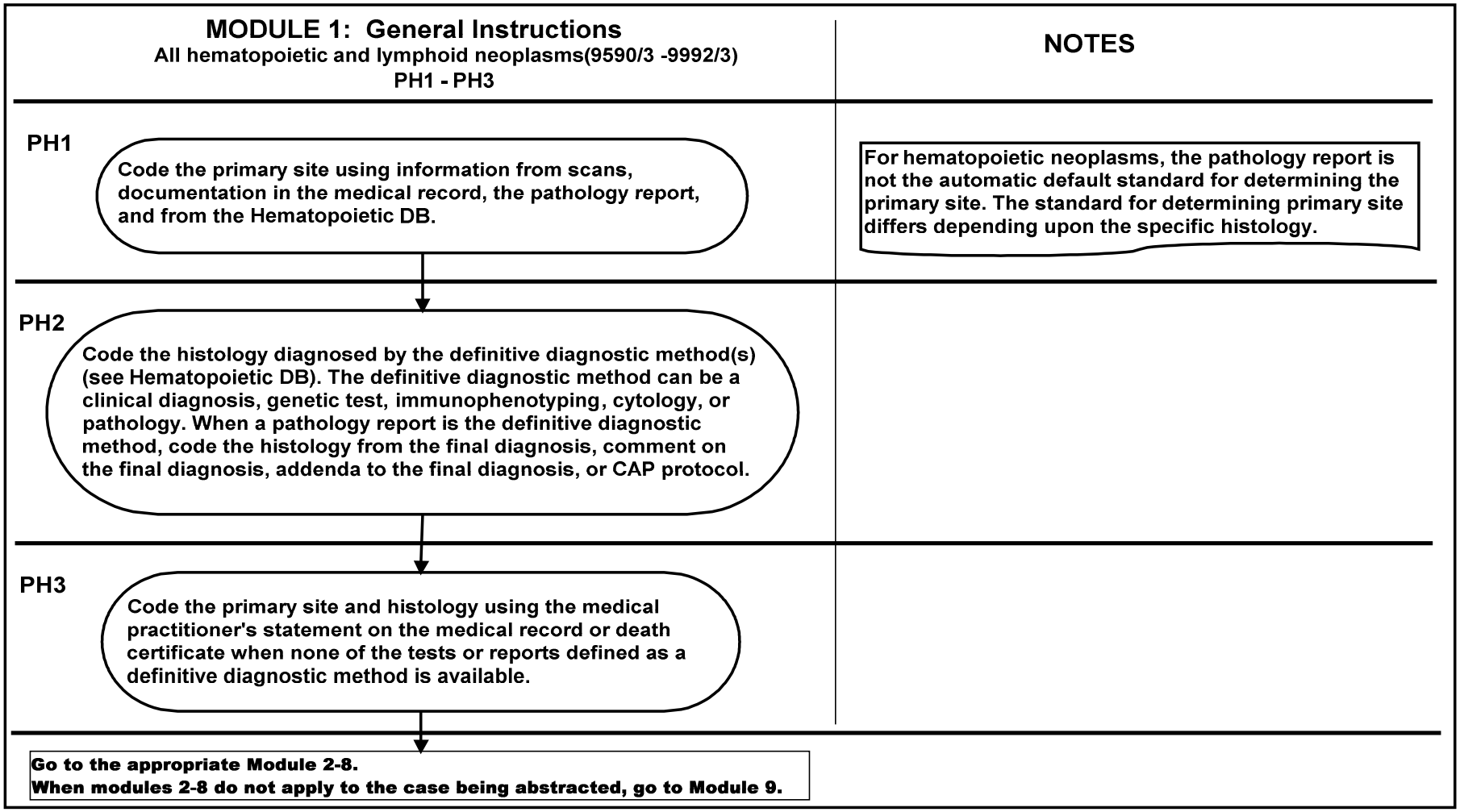
Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Note 1: Use the Primary Site and Histology Rules **before** using the Hematopoietic DB

Note 2: The primary site and histology coding rules are divided into nine modules. Each module covers a group of related hematopoietic or lymphoid **neoplasms.** However, a specific histology may be covered in more than one module.

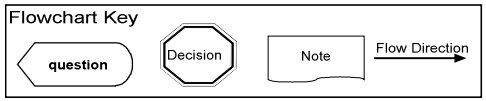
Note 3: The **modules** are **not hierarchical**, but the **rules** within each module **are** in **hierarchical** order. Apply the rules **within each module** in order. **Stop** at the first rule that applies.

Note 4: Apply rules in Module 1 first. Then go to the **first module** that **applies** to the case you are abstracting. If the situation in your case is not covered in that module **continue** on **as directed** after the last rule in the module.



Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules **before** using the Hematopoietic DB.



MODULE 2: Plasma Cell Neoplasms (9731/3, 9732/3, 9734/3) PH4-PH8

PH4

Does any of the following plasmacytoma occur in a site other than bone?
- **Plasmacytoma** or
- **Extraosseous (extramedullary) plasmacytoma** or
- **Solitary plasmacytoma** or
- **Multiple plasmacytomas** or
- **Multiple extraosseous (extramedullary) plasmacytomas**?

YES → **PRIMARY SITE:** Code to the **site of origin** (lymph node region(s), tissue, or organ)

HISTOLOGY: Code to extra-medullary plasmacytoma (9734/3)

NOTES and EXAMPLES

1. Extramedullary and extraosseous mean "not occurring in bone."

2. 80% of extramedullary plasmacytomas occur in the upper respiratory tract (oropharynx, nasopharynx, sinuses, and larynx) although they may occur in numerous other sites including the GI tract, lymph nodes, bladder, CNS, breast, thyroid, testis, parotid, and skin.

3. Do **not** code to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

Example 1: Pathology reports a solitary plasmacytoma wrapped around L4 vertebrae, no invasion of vertebrae. Code the primary site as soft tissue (C496) and the histology 9734/3.

Example 2: Scan shows two plasmacytomas in the nasopharyngeal wall. Biopsy confirms plasmacytoma. Code the primary site nasopharynx (C119) and the histology 9734/3.

NO ↓

PH5

Is the diagnosis:
- **Plasma cell neoplasm** or
- **Solitary plasmacytoma** or
- **Solitary plasmacytoma of bone** or
- **Solitary medullary plasmacytoma** or
- **Multiple plasmacytomas** or
- **Multiple plasmacytomas of bone** or
- **Multiple medullary plasmacytoma**?

YES → **PRIMARY SITE:** Code to the **specific bone (C400-C419)** where the plasmacytoma originated

HISTOLOGY: Code to solitary plasmacytoma of bone (9731/3)

NOTES and EXAMPLES

1. The most common sites are bones with active bone marrow hematopoiesis; in order of frequency these include vertebrae, ribs, skull, pelvis, femur, clavicle, and scapula.

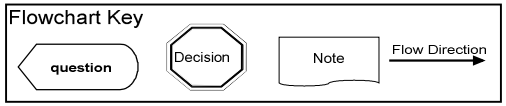
2. Do **not** code primary site to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

NO ↓

Next Page

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules **before** using the Hematopoietic DB.



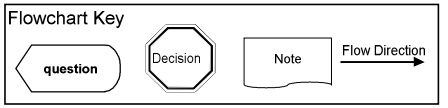
MODULE 2: Plasma Cell Neoplasms
(9731/3, 9732/3, 9734/3)
PH4-PH8

	PRIMARY SITE	HISTOLOGY	NOTES and EXAMPLES
PH6 Is there only information that the patient had a **plasmacytoma** or a **solitary plasmacytoma**? YES	Code to **unknown (C809)**	Code to solitary plasmacytoma of bone (9731/3)	***Example:*** Death-certificate-only case with cause of death listed as plasmacytoma.
NO			
PH7 Is the **clinical** diagnosis plasma cell myeloma/multiple myeloma and there is no documentation of bone marrow biopsy or the results of the bone marrow biopsy are unknown or unavailable? YES	Code to **bone marrow (C421)**	Code to plasma cell myeloma/ **multiple myeloma** (9732/3)	***Example:*** Death-certificate-only case with underlying cause of death listed as multiple myeloma. A clinical diagnosis of multiple myeloma may be based on myloidosis with associated renal impairment, anemia, and/or hypercalcemia supported by radiologic evidence of multiple lytic bone lesions.
NO			
PH8 Is the diagnosis **smoldering myeloma, indolent myeloma, evolving myeloma, plasma cell myeloma,** or **multiple myeloma**? YES	Code to **bone marrow (C421)**	Code to plasma cell myeloma/ **multiple myeloma** (9732/3)	1. When the proportion of plasma cells in the bone marrow is 10% or greater, the diagnosis is multiple myeloma. 2. A medical record may have multiple bone marrow biopsies. If any one of the biopsies is positive for multiple myeloma, code the histology to multiple myeloma and the primary site to bone marrow. ***Example:*** Bone marrow biopsies: Biopsy 1: Negative. Biopsy 2: Multiple myeloma with bone marrow showing 18% plasma cells. Code the primary site bone marrow C421 and the histology 9732/3.
NO			

When this module does not apply to the plasma cell neoplasm (9731/3, 9732/3, 9734/3) case being abstracted, go to Module 8.

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

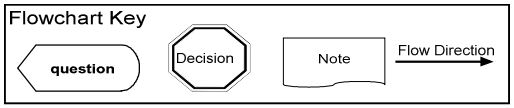
Use the Primary Site and Histology Rules **before** using the Hematopoietic DB.



Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma
Note 2: Commonly lymphoma originates in lymph node region(s), tissue, or organ(s) although it will metastasize to the bone marrow when the disease is stage IV or disseminated
Note 3 Commonly leukemia originates in the bone marrow

MODULE 3: Lymphoma/Leukemia (Specific neoplasms that can manifest as leukemia or as lymphoma) (9670/3, 9687/3, 9727/3, 9728/3, 9729/3, 9823/3, 9826/3, 9835/3, 9836/3, 9837/3) PH9-PH12	PRIMARY SITE	HISTOLOGY	NOTES and EXAMPLES
PH9 Is the diagnosis B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **(BCCLL/SLL)** and **peripheral blood** is involved (the **bone marrow** may also be involved)? YES	Code to **bone marrow (C421)**	Code to B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **(BCCLL/CLL) (9823/3)**	1. Peripheral blood involvement requires repeated CBCs with absolute lymphocyte count >5000 on repeated measures or flow cytometry that documents a clonal B-cell population in the bone marrow. 2. Leukemic BCCLL will always have peripheral blood involvement. The bone marrow may or may not be involved. In later stages of the disease there may be involvement of lymph nodes, liver and spleen. 3. Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.
NO			
PH10 Is the diagnosis B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **AND** you **cannot verify** that the disease originated in the bone marrow? YES	Code to **site of origin** (lymph node region(s), tissue, or organ)	Code to **small B lymphocytic lymphoma (9670/3)**	1. Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. 2. See Appendix C for help in identifying lymph node regions and codes. 3. In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow. 4. Small lymphocytic lymphoma is characterized by negative peripheral blood involvement (an absolute lymphocyte count <=5000 on repeated CBCs).
NO			
Next Page			

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm



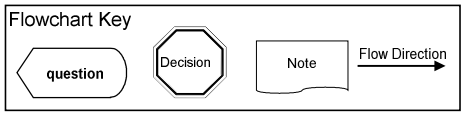
Use the Primary Site and Histology Rules before using the Hematopoietic DB.

Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma
Note 2: Commonly lymphoma originates in lymph node region(s), tissue, or organ(s) although it will metastasize to the bone marrow when the disease is stage IV or disseminated
Note 3: Commonly leukemia originates in the bone marrow

MODULE 3: Lymphoma/Leukemia (Specific neoplasms that can manifest as leukemia or as lymphoma) (9670/3, 9687/3, 9727/3, 9728/3, 9729/3, 9823/3, 9826/3, 9835/3,9836/3, 9837/3) PH9-PH12	PRIMARY SITE	HISTOLOGY	NOTES and EXAMPLES
PH11 Is the **only** involvement **bone marrow AND** the diagnosis is: • Burkitt cell leukemia (9826/3) or • Precursor cell lymphoblastic leukemia, NOS (9835/3) or • Precursor B-cell lymphoblastic leukemia/lymphoma (9836/3) or • T lymphoblastic leukemia/lymphoma (9837/3)? YES	Code to **bone marrow (C421)**	Code to **the respective histology from the list**	1. Leukemia most commonly originates in the bone marrow. When only the bone marrow is involved, code as leukemia. 2. Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.
NO			
PH12 Is there involvement of lymph nodes, tissue, or organ(s) **AND** the diagnosis is Burkitt **lymphoma/leukemia or**: • Burkitt lymphoma, NOS (9687/3) or • Blastic plasmacytoid dendritic cell neoplasm (9727/3) (previously called precursor cell lymphoblastic lymphoma, NOS) or • Precursor B-cell lymphoblastic lymphoma (9728/3) or • Precursor T-cell lymphoblastic lymphoma (9729/3)? YES	Code to the **site of origin** (lymph node region(s), tissue, or organ)	Code **to the respective histology from the list**	1. Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. 2. See Appendix C for help in identifying lymph node regions, and codes. 3. In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow.
NO			
When this module does not apply to the case being abstracted, go to Module 8.			

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules before using the Hematopoietic DB.



MODULE 4: Preleukemia, Smoldering Leukemia and Myelodysplastic Syndrome (9989/3)

PRIMARY SITE	HISTOLOGY	NOTES and EXAMPLES

PH13

Is the diagnosis **preleukemia, smoldering leukemia, or myelodysplastic syndrome?**

YES → Code to **bone marrow (C421)** → Code to **myelo-dysplastic syndrome (9989/3)**

NO → **When this module does not apply to the case being abstracted, go to Module 8.**

MODULE 5: Myeloid Neoplasms (9861/3, 9930/3)

PH14-PH15

PRIMARY SITE	HISTOLOGY	NOTES and EXAMPLES

PH14

Is the diagnosis **myeloid neoplasm** or **acute myeloid leukemia, NOS AND** the involvement is limited to bone marrow?

YES → Code to **bone marrow (C421)** → Code to **acute myeloid leukemia, NOS 9861/3**

Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

NO

PH15

Is the diagnosis **myeloid neoplasm** or **myeloid sarcoma** AND the neoplasm originates in a site other than bone marrow?

YES → Code to **site of origin** (lymph node region(s), tissue, or organ) → Code to **myeloid sarcoma (9930/3)**

1. Most common sites are skin, lymph node(s), GI tract, bone, soft tissue, and testis. This neoplasm however, can occur in almost every site of the body other than bone. It **does not** originate in bone marrow.

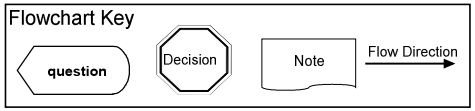
2. See Appendix C for help in identifying lymph node names, chains, and codes.

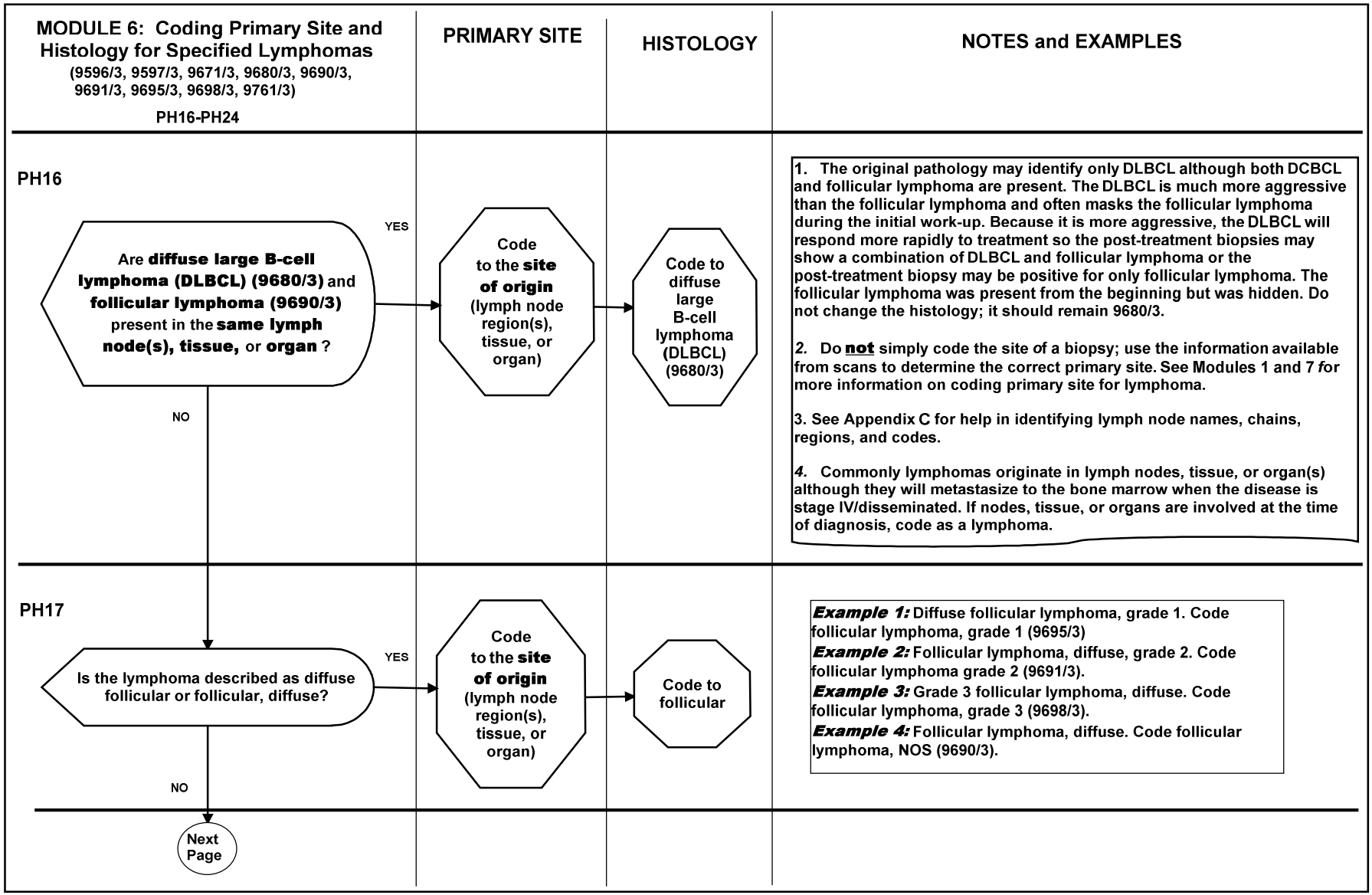
NO

For information on coding primary site for lymphomas go to Modules 1 and 7.
When this module does not apply to the case being abstracted, go to Module 8.

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

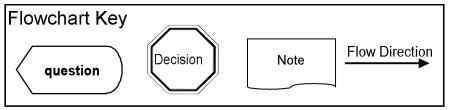
Use the Primary Site and Histology Rules before using the Hematopoietic DB.

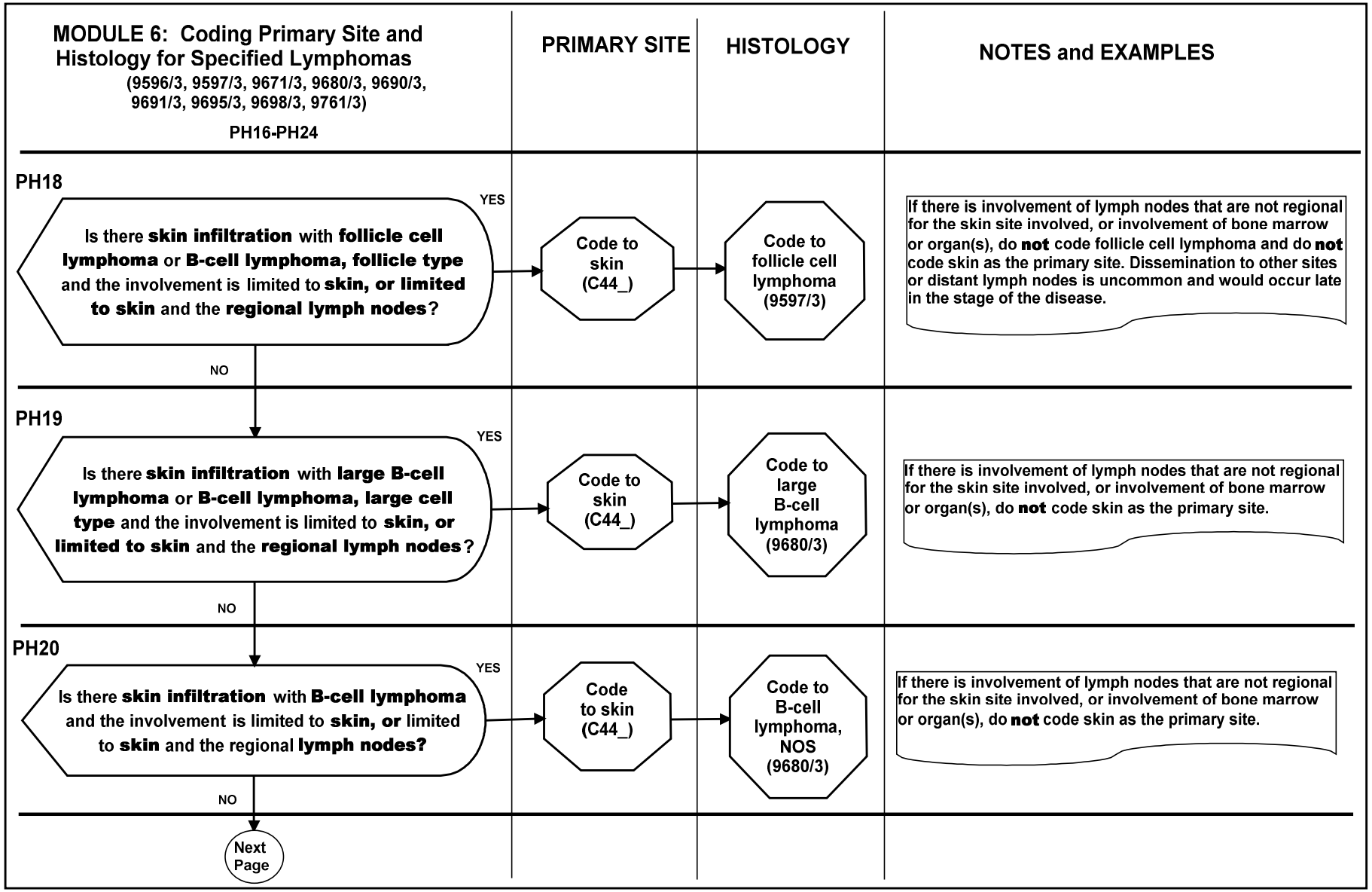




Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

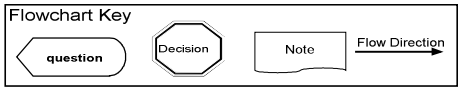
Use the Primary Site and Histology Rules before using the Hematopoietic DB.





Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

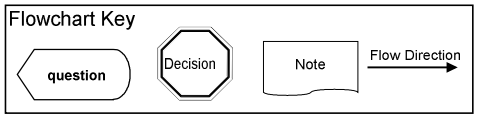
Use the Primary Site and Histology Rules before using the Hematopoietic DB.

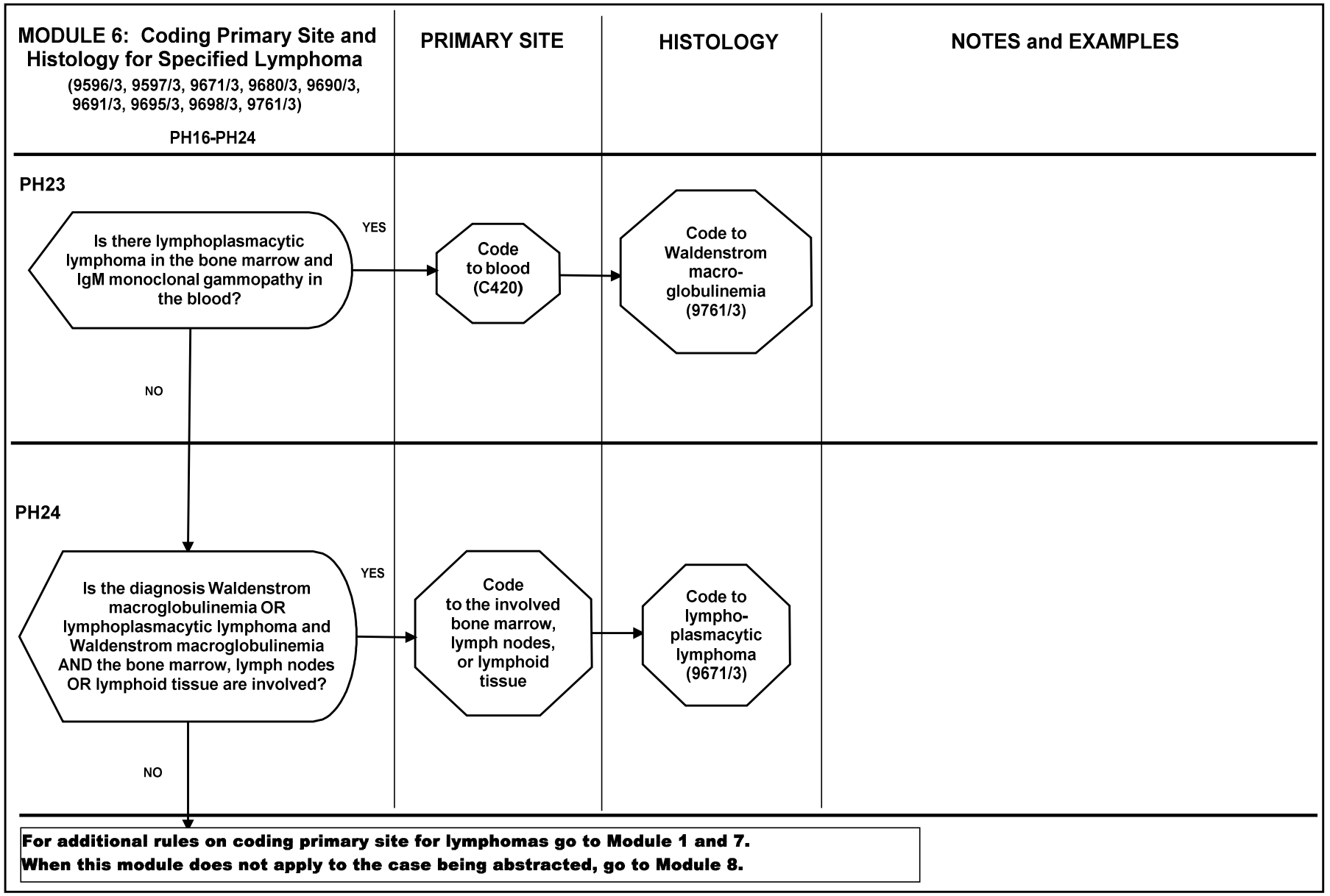


MODULE 6: Coding Primary Site and Histology for Specified Lymphomas (9596/3, 9597/3, 9671/3, 9680/3, 9690/3, 9691/3, 9695/3, 9698/3, 9761/3) PH16-PH24	PRIMARY SITE	HISTOLOGY	NOTES and EXAMPLES
PH21 Are **both** non-Hodgkin lymphoma and Hodgkin lymphoma **present in the same lymph node region(s), tissue,** or **organ** ? YES → Code to the **site of origin** (lymph node region(s), tissue, or organ) → Code to **composite lymphoma (9596/3)** NO ↓			1. Use the composite lymphoma code when: ● Both NHL and HL are present in one lymph node or multiple lymph nodes in one lymph node region. ● Both NHL and HL are present in multiple lymph nodes in one lymph node region or several lymph node regions as defined by ICD-O-3. i.e.: NHL and HL present in superior hilum and superior rectal lymph nodes. ○ Assume all lymph nodes are involved with both NHL and HL even though only one lymph node is biopsied. 2. Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. 3. See Appendix C for help in identifying lymph node names, chains, and codes. 4. Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma 5. Do **not** use the composite lymphoma code 9596/3 when: ● NHL is present in one lymph node region and HL is present in another lymph node region i.e.: NHL in cervical lymph node(s) and HL in inguinal lymph node(s) ● NHL in liver and HL in intra-thoracic lymph nodes
PH22 Are two or more **non-Hodgkin lymphomas** present in the same lymph node(s), tissue, or organ? YES → Code to the **site of origin** (lymph node region(s), tissue, or organ) → Code to the **numerically highest ICD-O-3 code** NO ↓			1. Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. 2. See Appendix C for help in identifying lymph node names, chains, regions, and codes. 3. Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma. 4. This rule does not apply when NHL is present in one node and a different NHL histology is present if: ● Thymic extranodal marginal-zone B-cell lymphoma is present in the thymus and diffuse large B-cell lymphoma in the hilar lymph nodes. ● B-cell lymphoma is present in the intrathoracic lymph nodes and peripheral T-cell NHL in the liver.
Next Page			

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

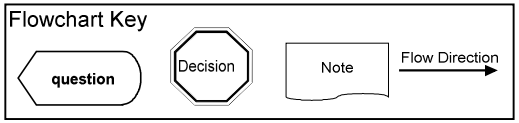
Use the Primary Site and Histology Rules before using the Hematopoietic DB.

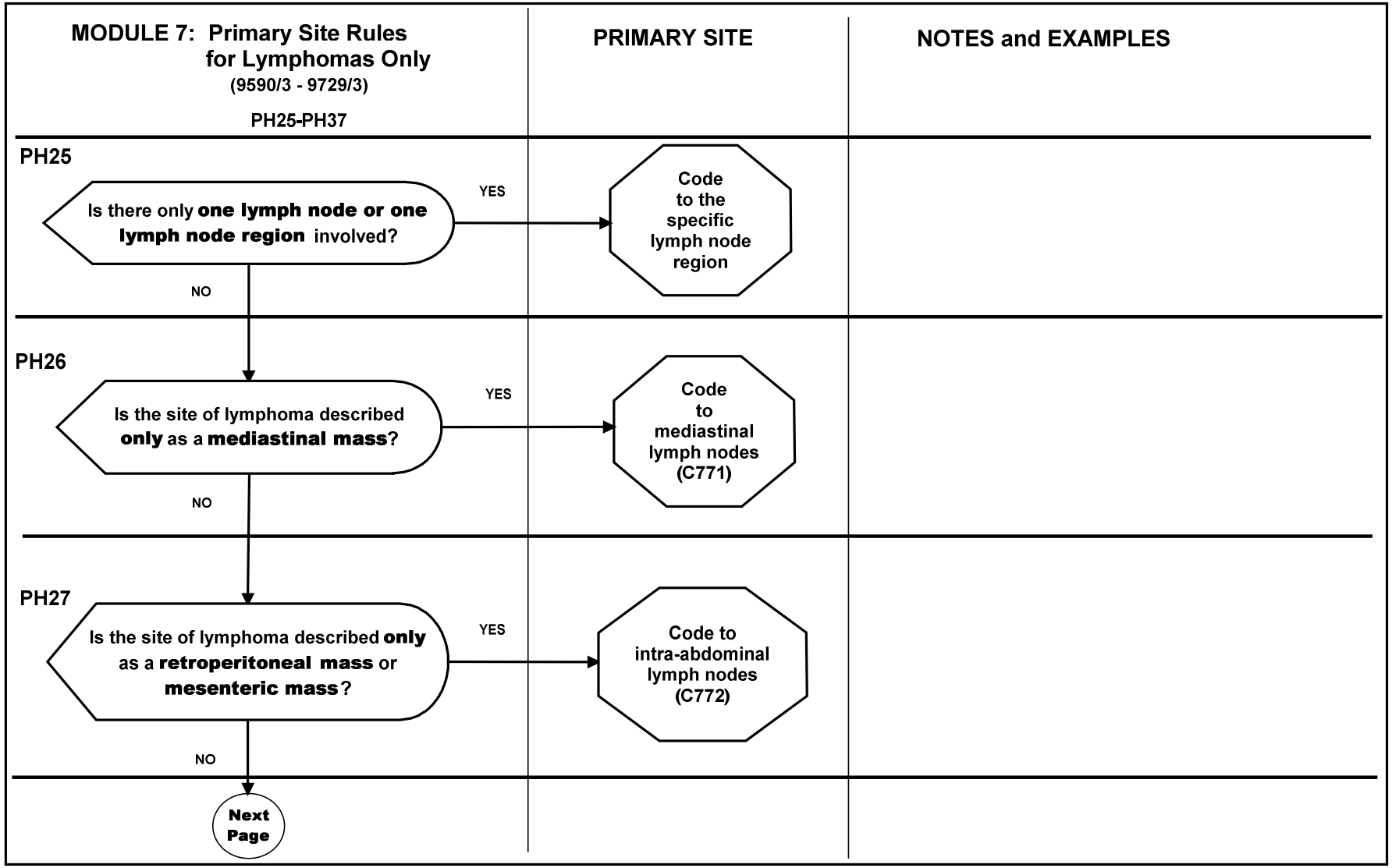




Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

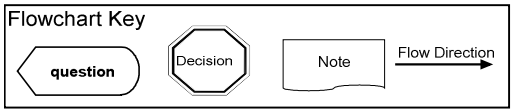
Use the Primary Site and Histology Rules before using the Hematopoietic DB.





Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules before using the Hematopoietic DB.



MODULE 7: Primary Site Rules for Lymphomas Only
(9590/3 - 9729/3)

PH25-PH37

PRIMARY SITE

NOTES and EXAMPLES

PH28

Is the site of lymphoma described only as an inguinal mass?

YES → **Code to inguinal lymph nodes (C774)**

NO

PH29

Are multiple lymph node chains within the same region (as defined by ICD-O-3) involved?

YES → **Code to the specific lymph node region**

Example 1: Code intra-abdominal lymph nodes (C772) when there is involvement of hepatic (C772) and para-aortic lymph node chains (C772).

Example 2: Code lymph nodes of head, face and neck (C770) when there is involvement of cervical (C770) and mandibular (C770) lymph node chains.

Example 3: Code mediastinal lymph nodes (C771) when bilateral mediastinal lymph nodes are involved.

NO

PH30

Are multiple lymph node regions (C778) defined by ICD-O-3 involved and it is not possible to identify the lymph node region where the lymphoma originated?

YES → **Code to multiple lymph node regions (C778), NOS**

1. Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Module 1 for more information on coding primary site for lymphoma.

2. See Appendix C for help in identifying lymph node names, chains, regions, and codes.

Example 1: Cervical and intrathoracic lymph nodes involved with B-cell lymphoma.

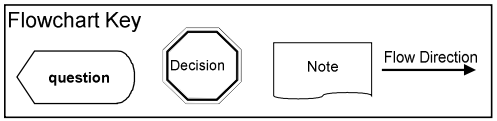
Example 2: CT scans showed involvement of the cervical lymph nodes (C770) and the mediastinal lymph nodes (C771). No additional involvement was identified during the work-up. Biopsy of a cervical lymph node confirmed lymphoma. Code the primary site to lymph nodes of multiple regions (C778).

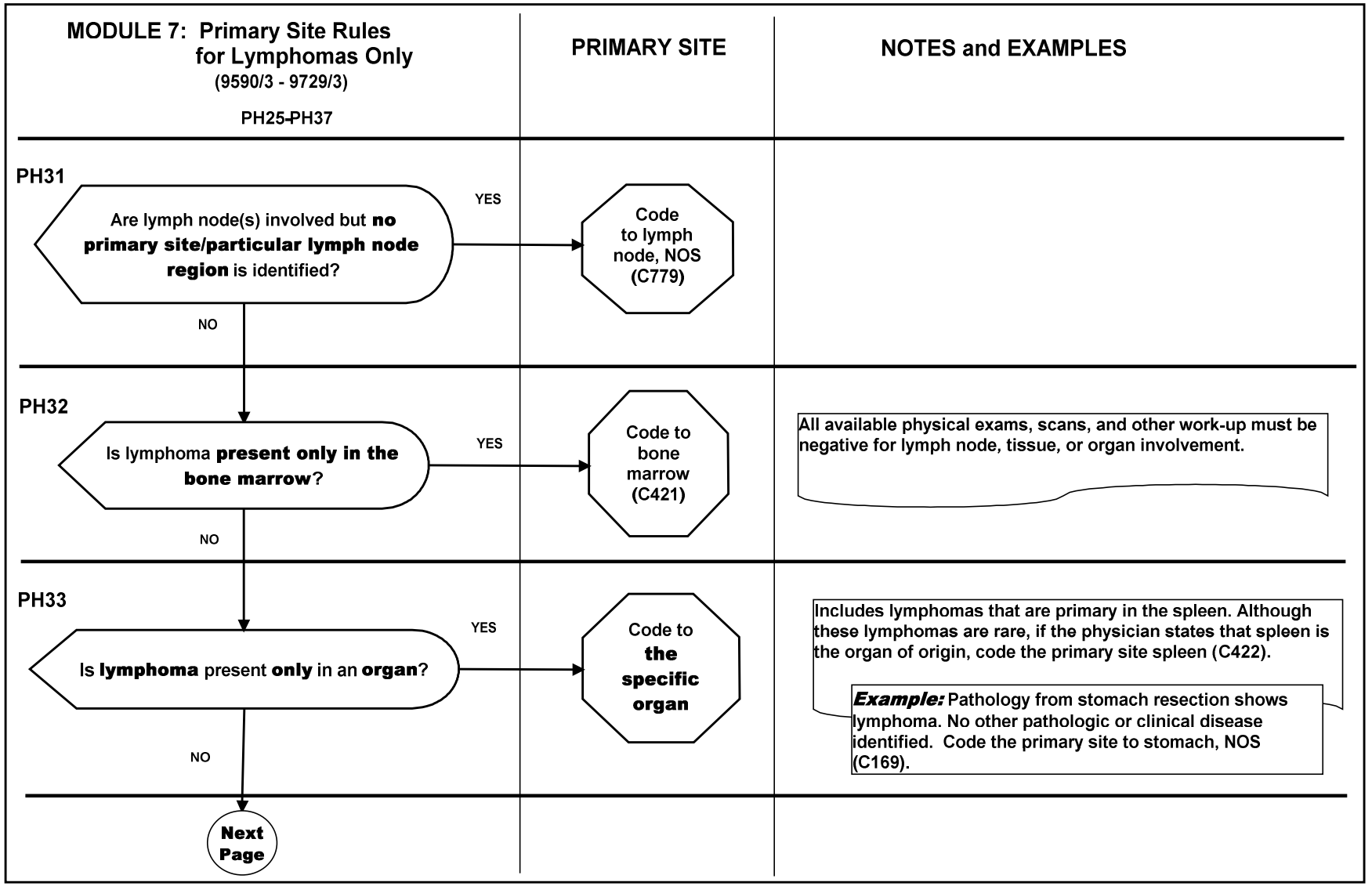
NO

Next Page

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

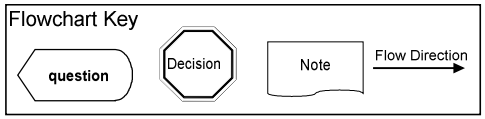
Use the Primary Site and Histology Rules **before** using the Hematopoietic DB.





Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules **before** using the Hematopoietic DB.



MODULE 7: Primary Site Rules for Lymphomas Only

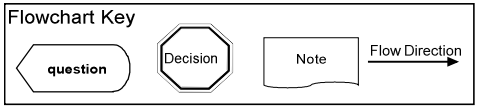
(9590/3 - 9729/3)

PH25-PH37

	PRIMARY SITE	NOTES and EXAMPLES
PH34 Is there **proof of extension from the regional lymph nodes** into the organ?	YES → Code to the lymph node region as defined by ICD-O-3	***Example:*** Patient presents with abdominal adenopathy. Surgical exploration documents direct invasion of the stomach from the regional lymph nodes. Code abdominal lymph nodes (C772).
NO		
PH35 Is **lymphoma** present in an **organ** and that organ's **regional lymph nodes**?	YES → Code to the organ	Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes. ***Example 1:*** Lymphoma is present in the kidney and peri-renal lymph nodes. Code the primary site to kidney (C649). ***Example 2:*** Lymphoma is present in the stomach and the gastric lymph nodes. Code the primary site to stomach, NOS (C169). ***Example 3:*** Lymphoma is present in the spleen and the splenic lymph nodes. Code the primary site spleen (C422).
NO		
PH36 Is **lymphoma** present in an **organ(s)** and **lymph nodes** that are **not regional** for that organ and the **origin cannot be determined** even after consulting the physician?	YES → Code to lymph nodes, NOS (C779)	1. Lymphoma can spread from organs to **regional** lymph nodes, but does **not** spread from the organ directly to **distant** lymph nodes. ***Example:*** The patient has positive mediastinal (C771) and cervical (C770) lymph nodes and involvement of the stomach (C169). No further information can be obtained. Code to lymph node, NOS (C779). 2. Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes. 3. See Appendix C for help in identifying lymph node names, chains, regions, and codes.
NO		
Next Page		

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules **before** using the Hematopoietic DB.



MODULE 7: Primary Site Rules for Lymphomas Only

(9590/3-9729/3)

PH25-PH37

PRIMARY SITE

NOTES and EXAMPLES

PH37

Is there **no evidence** of **lymphoma** in **lymph nodes** AND the physician documents in the medical record that he/she suspects that the **lymphoma originates in an organ(s)**? See ICD-O-3 Rule D.

YES → Code to unknown primary site (C809)

NO

When this module does not apply to the case being abstracted, go to Module 8

MODULE 8: Histology Rules Only

(9590/3 - 9992/3)

PH38 - PH39

HISTOLOGY

NOTES and EXAMPLES

PH38

Is the diagnosis:

- **One non-specific histology AND**
- **Two or more specific** histologies **AND**
- The hematopoietic DB calculator documents the specific histology and NOS are the **same primary AND**
- No further information is available?

YES → Code the non-specific (NOS) histology

1. Use Appendix E: Histology "NOS" and More Specific Tables to identify the NOS histologies.

2. Use the Hematopoietic DB multiple primary calculator to confirm that the NOS and specific histology are the same primary.

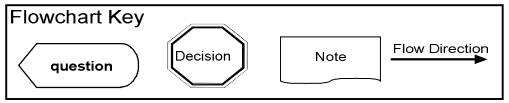
Example: The diagnosis is myeloproliferative disorder, NOS (9960/3), polycythemia vera (9950/3), essential thrombocythemia (9962/3).. The Hematopoietic DB multiple primary calculator shows myeloproliferative disorder and polycythemia vera are the same primary. The multiple primary calculator also shows myeloproliferative disorder and essential thrombocythemia are the same primary. Follow-back produces no additional information. Code the histology myeloproliferative disorder, NOS (9960/3).

NO

Next Page

Primary Site and Histology Coding Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

Use the Primary Site and Histology Rules before using the Hematopoietic DB.



MODULE 8: Histology Rules Only

All hematopoietic and lymphoid neoplasms (9590/3 - 9992/3)

PH38 - PH39

	HISTOLOGY	NOTES and EXAMPLES
PH39 Is the diagnosis: ● **One non-specific(NOS) histology AND** ● **One specific** histology **AND** ● The hematopoietic DB calculator documents the specific histology and NOS are the **same primary** YES	Code the **specific** histology	1. Use Appendix E: Histology "NOS" Tables to identify the NOS histologies. 2. Use the Hematopoietic DB multiple primary calculator to confirm that the NOS and specific histology are the same primary.

NO

When this module does not apply to the case being abstracted, go to Module 9.

MODULE 9: Default Rules

All hematopoietic and lymphoid neoplasms (9590/3 - 9992/3)

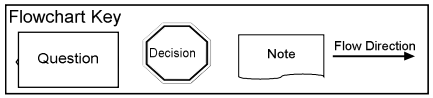
PH40-PH41

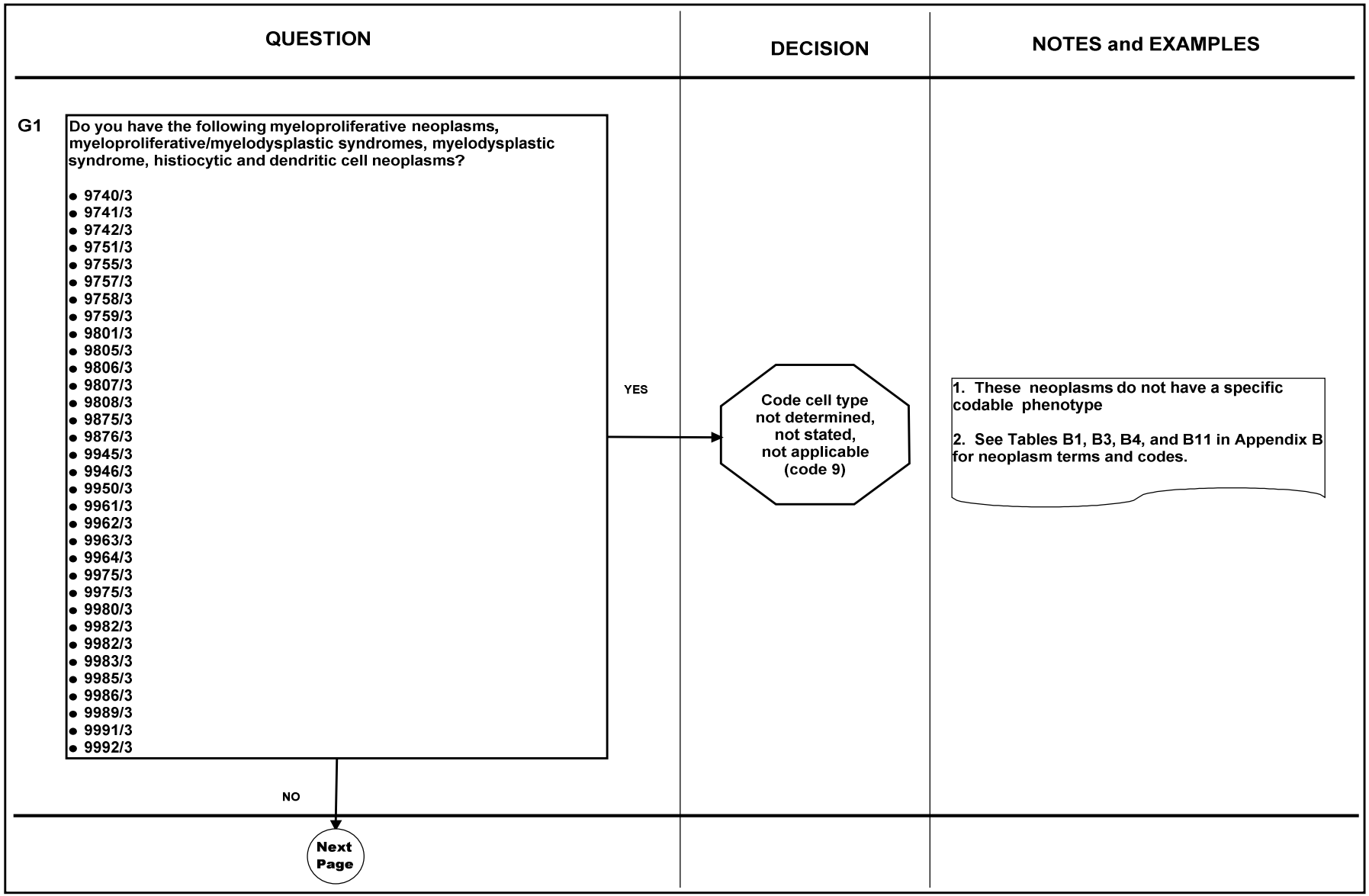
	HISTOLOGY	NOTES and EXAMPLES
PH40 Is it impossible to apply rules PH1-PH39 to the case being abstracted? YES	Use the **Hematopoietic DB** to determine the primary site and histology	
NO		
PH41 Is it impossible to determine code using the Hematopoietic DB? YES	Code the histology with the **numerically higher** ICD-O-3 code	
NO		

This is the end of rules for coding primary site and histology

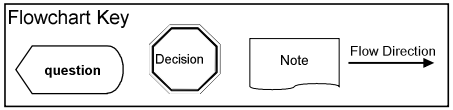
Grade of Tumor Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

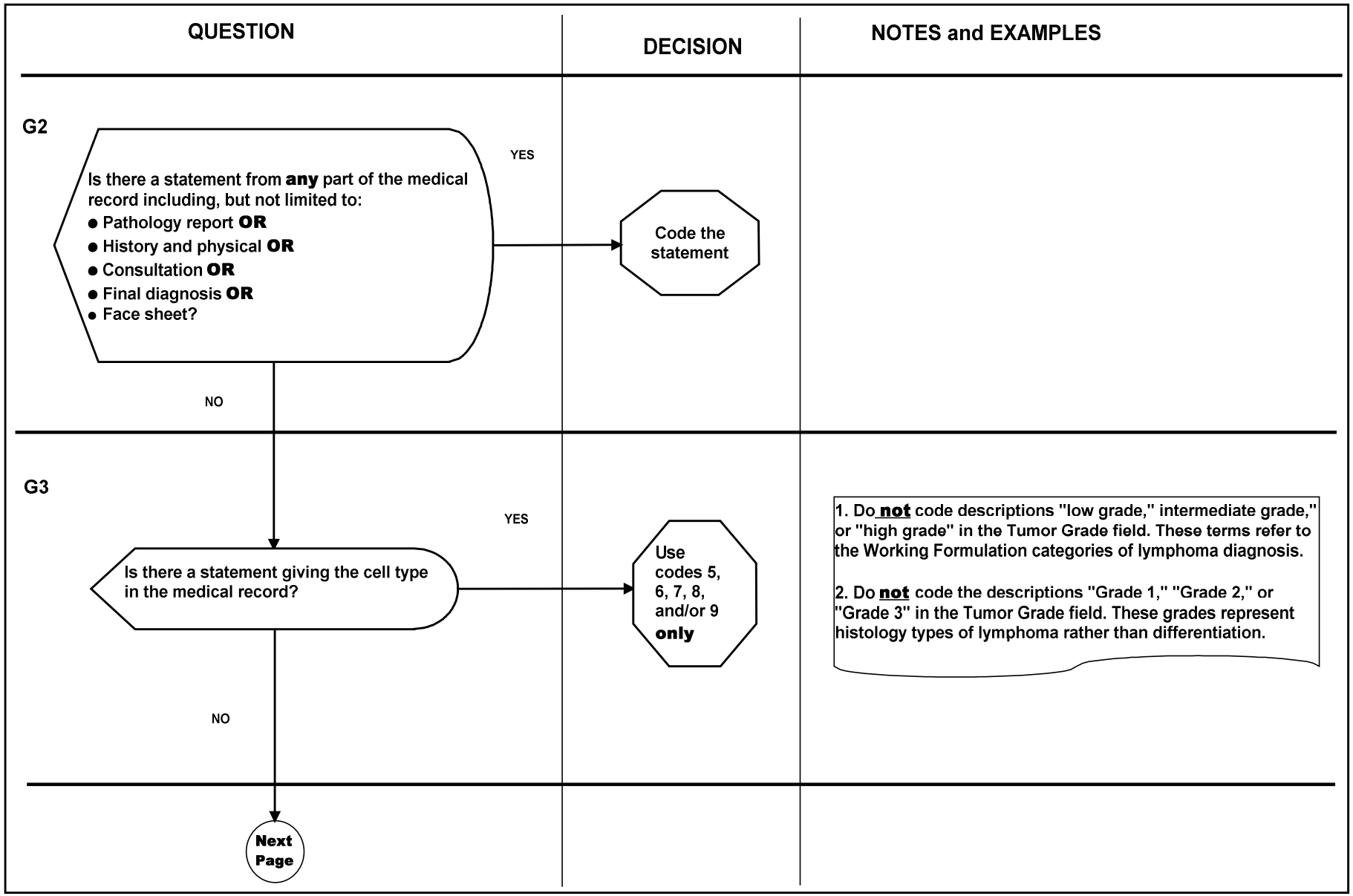
Note 1: Use the Grade of Tumor Rules (G1-G11) to assign the correct code in the grade field
Note 2: Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade



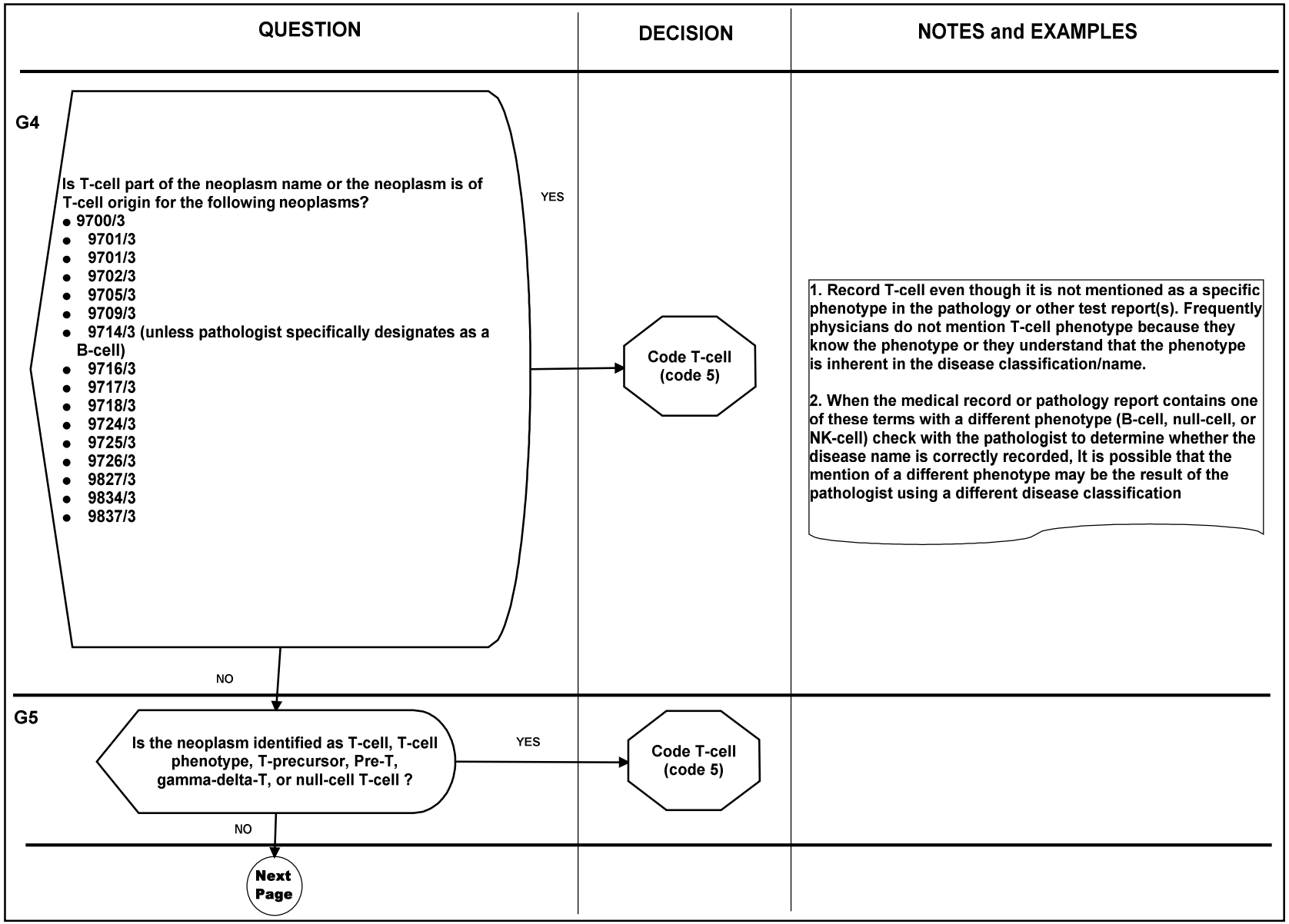


Grade of Tumor Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm

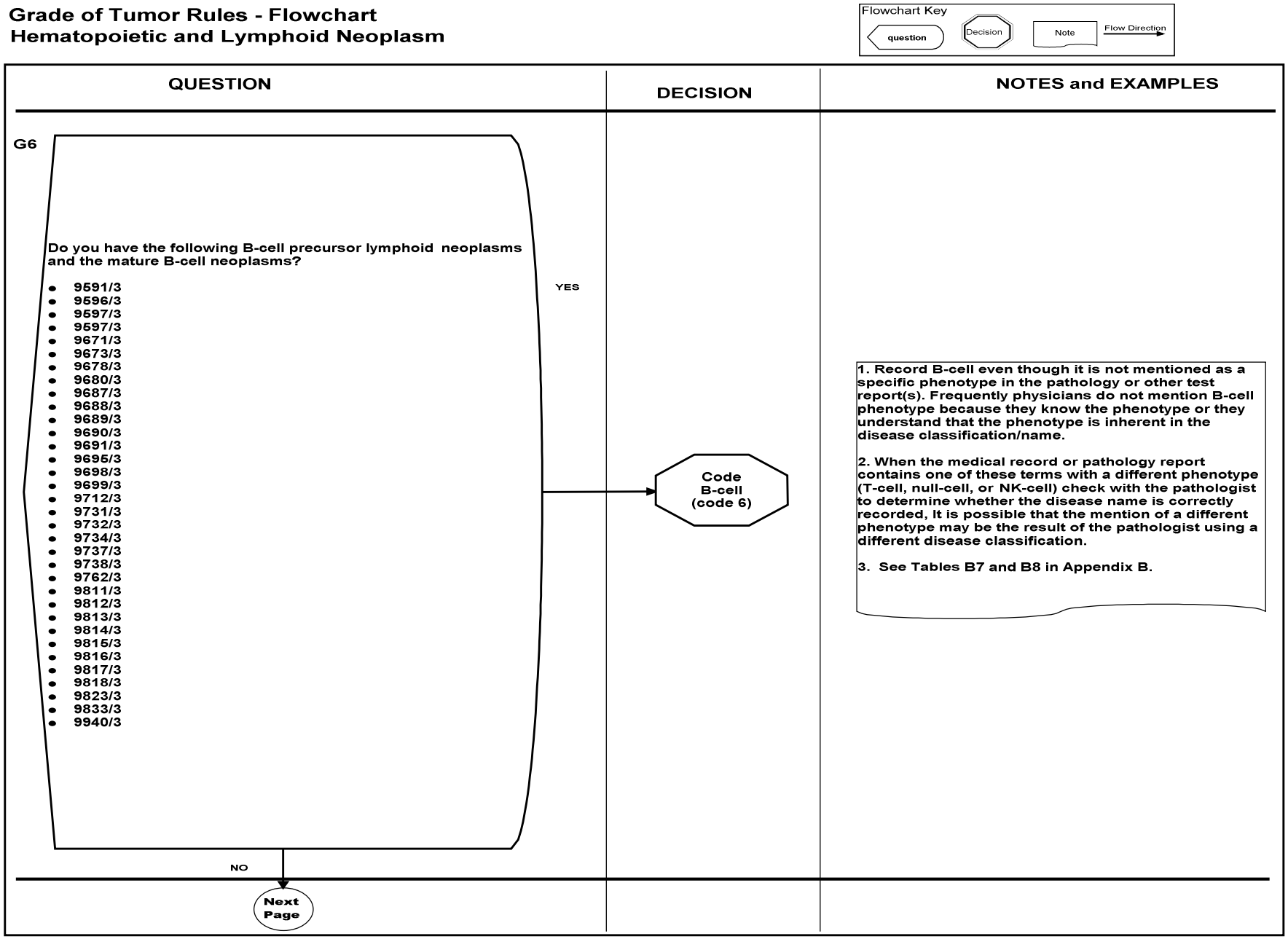




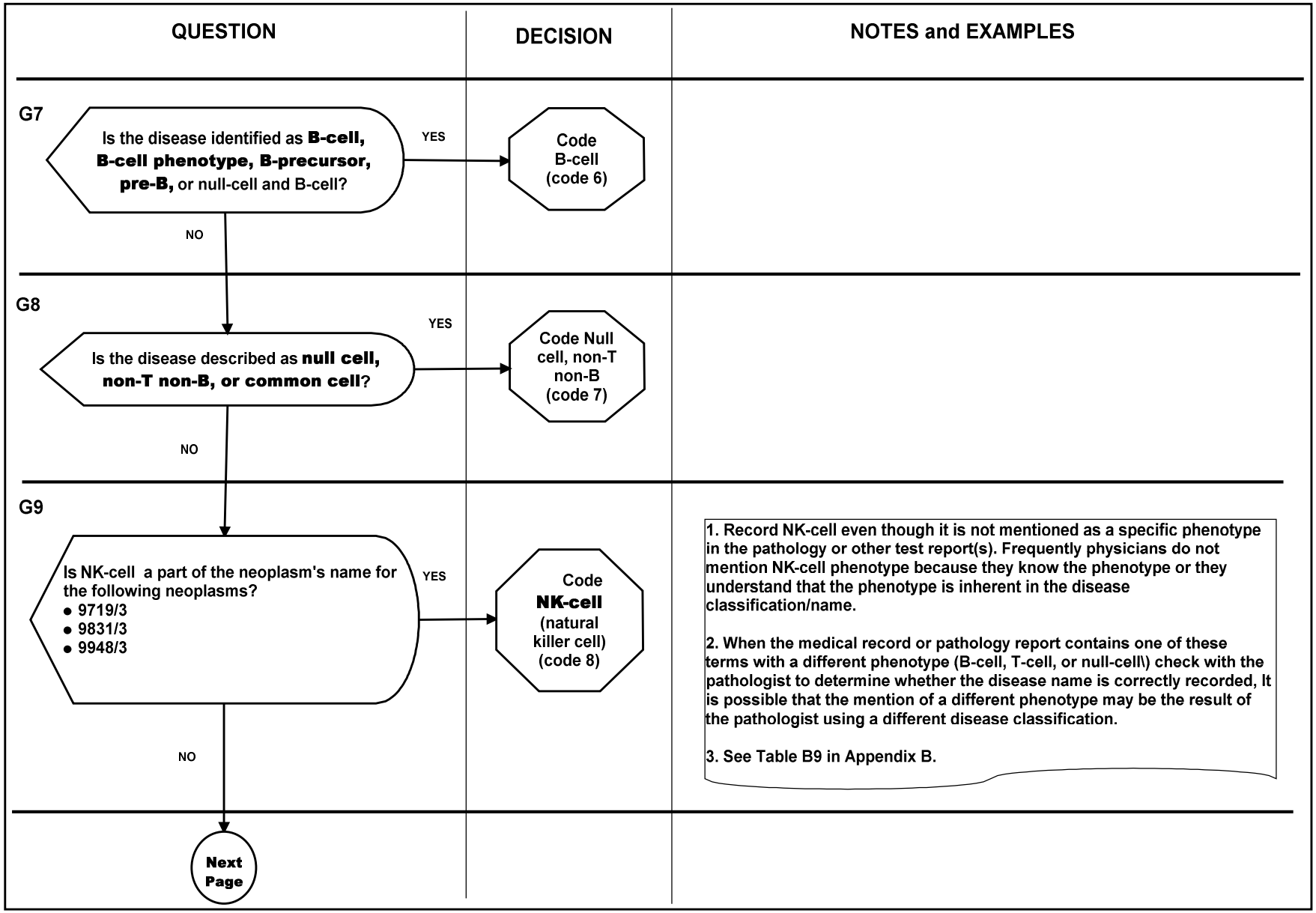
Grade of Tumor Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm



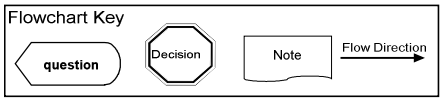
Grade of Tumor Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm



Grade of Tumor Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm



Grade of Tumor Rules - Flowchart
Hematopoietic and Lymphoid Neoplasm



QUESTION	DECISION	NOTES and EXAMPLES
G10		
Is the disease described as **NK cell, natural killer cell, nasal NK/T-cell** lymphoma, or null-cell and NK cell?	YES → Code Natural Killer (NK) cell (code 8)	
NO		
G11		
Is there any of the following? ● There is **no statement describing the cell type** OR ● **The cell type is described as combined T** AND **B cell** OR ● The cell type is described as **combined B** AND **NK cell**	YES → Code cell type not determined, not stated, not applicable (code 9)	There is a new site-specific factor to collect combination cell types for a hematopoietic or lymphatic neoplasm in the Collaborative Stage Data Collection System Version 2.
NO		
This is the end of Grade of Tumor Rules		

Multiple Primary Rules Matrix Format

Note 1: Use Multiple Primary Rules M1 through M12 **before** using the Hematopoietic DB.
Note 2: The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules M7-M12.

Rule	Histology	Number of Primaries	Examples/Comments
M1	Minimal information available, such as DCO case or a pathology only case	Single*	
M2	Single histology	Single*	***Example 1***: The diagnosis is multiple myeloma (9732/3). Abstract as a single primary. ***Example 2***: Multiple extraosseous plasmacytomas (9734/3) are present in the oropharynx. Abstract as a single primary. ***Example 3***: A single histology diagnosed by the definitive diagnostic method as defined in the Hematopoietic DB; **for example**. The patient had several provisional diagnoses but the definitive diagnostic method identifies a single histology. Abstract as a single primary.
M3	**Two or more types of non-Hodgkin lymphoma** are present in the **same anatomic location(s)**, such as • One lymph node **OR** • One organ **OR** • One tissue	Single*	***Example***: Biopsy of cervical lymph node shows **two** different non-Hodgkin lymphomas. Abstract as a single primary. ***Note 1***: Do **NOT** use this rule for cutaneous lymphomas**.** ***Note 2***: When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for information on lymph node codes and regions. ***Note 3***: When the disease is in a more advanced stage, both non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3 or in an organ and that organ's regional lymph nodes or in multiple organs. • Although both non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination non-Hodgkin lymphoma, assume that all of the nodes, tissue, and/or organs are involved with the combination of non-Hodgkin lymphomas. ***Note 4***: Do **not** query the Hematopoietic DB in this situation.
M4	Both **Hodgkin and non-Hodgkin lymphoma** are present in the **same** anatomic **location(s)**. Hodgkin and non-Hodgkin may be present in • One lymph node **OR** • One organ **OR** • One tissue	Single*	***Example***: Biopsy of cervical lymph node shows Hodgkin and non-Hodgkin lymphomas. Abstract as a single primary. ***Note 1***: When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for lymph node codes and regions. ***Note 2***: When the disease is in a more advanced stage, both Hodgkin and non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3 codes or in an organ and that organ's regional lymph nodes or in multiple organs. • Although both Hodgkin and non-Hodgkin lymphomas must be present in each of

Rule	Histology	Number of Primaries	Examples/Comments
			the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination Hodgkin and non-Hodgkin lymphomas, assume that all of the nodes, tissue, and/or organs are involved with the combination of Hodgkin and non-Hodgkin lymphomas. ***Note 3***: Do not query the Hematopoietic DB in this situation.
M5	• Hodgkin lymphoma in **one node** and non-Hodgkin lymphoma in a **different node** ***Note***: The involved nodes may be in the same lymph node region as defined by ICD-O-3 or in different lymph node regions as defined by ICD-O-3. See Appendix C. • Hodgkin lymphoma in **one organ** and non-Hodgkin lymphoma in a **different organ** • Hodgkin lymphoma in **tissue** and non-Hodgkin lymphoma **in different tissue**	Multiple**	***Example 1***: Patient is diagnosed with Hodgkin lymphoma in the cervical lymph nodes and also with non-Hodgkin lymphoma in the inguinal lymph nodes. Abstract as multiple primaries. ***Example 2***: Hodgkin lymphoma in thymus and non-Hodgkin lymphoma in the tonsil. Abstract as multiple primaries. ***Example 3***: Hodgkin lymphoma in the brain and non-Hodgkin lymphoma in the mediastinal lymph nodes. Abstract as multiple primaries
M6	• **More specific** histology diagnosed after an **NOS** • Hematopoietic DB Multiple Primary Calculator confirms that NOS and more specific histology are the same primary	Single*	***Note 1***: There are no time restrictions on these diagnoses; the interval between the NOS and the more specific histology does not affect this rule stating that the two neoplasms are a single primary. ***Note 2:*** The Hematopoietic DB will identify these histologies as a single primary
M7	• Both the **chronic and the acute phase** of the neoplasm are diagnosed **within 21 days AND** • **There is documentation** of **one positive** bone marrow biopsy	Single (Abstract the phase diagnosed by positive bone marrow)*	When these diagnoses happen within 21 days, it is highly possible that one diagnosis was provisional and the bone marrow identified the correct diagnosis.
M8	• Both the **chronic and the acute phase** of the neoplasm are diagnosed **within 21 days AND** • **There is documentation** of **two** bone marrow examinations, **one confirming the chronic neoplasm and another confirming the acute neoplasm**	Multiple** (both the chronic and the acute phase)	N/A
M9	• **Both the chronic and the acute phase** of the neoplasm are diagnosed **within 21 days AND** • There is **no available documentation**	Single* (the later diagnosis)	***Note 1***: The two diagnoses are likely the result of an ongoing diagnostic work-up. The later diagnosis is usually based on all of the test results. ***Note 2***: This rule applies if both neoplasms are diagnosed simultaneously (at the same time).

Rule	Histology	Number of Primaries	Examples/Comments
	on bone marrow biopsy		
M10	• **Originally diagnosed** in a **chronic** (less aggressive) phase **AND** • **Second diagnosis** of a blast or acute phase **more than 21 days** after the chronic diagnosis	Multiple**	***Note 1***: **This is a change from previous rules**. Use the Hematopoietic DB to determine multiple primaries when a transformation from the chronic to a blast or acute phase occurs. ***Note 2***: Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.
M11	• **Originally diagnosed** in blast or acute phase **AND** • Second diagnosis of- chronic phase (less aggressive) **AND** • **No confirmation** that patient was treated	Single* (acute phase)	***Note 1***: When these diagnoses happen **within 21 days**, it is highly possible that the first diagnosis of acute disease was a provisional diagnosis. ***Note 2***: When the subsequent diagnosis occurs more than 21 days after the original diagnosis of acute disease it is important to follow-back to obtain information on treatment or a subsequent bone marrow biopsy that negates the diagnosis of acute disease
M12	• **Originally diagnosed** in the blast or acute phase **AND** • **Reverts** to a chronic phase (less aggressive) after treatment	Multiple**	***Note 1***: Only abstract as a multiple primary when the patient has been treated for the acute disease. ***Note 2***: **This is a change from previous rules**. Use the Hematopoietic DB to determine multiple primaries when a transformation from the blast or acute phase to a chronic phase occurs. ***Note 3***: Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.
M13	Case does **not** meet the criteria in M1-M12	Use the Hematopoietic DB to determine the number of primaries	N/A

*Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.
**Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

Primary Site and Histology Coding Rules Matrix Format

Note 1: Use the Primary Site and Histology Rules before using the Hematopoietic DB
Note 2: The primary site and histology coding rules are divided into nine modules. Each **module** covers a group of **related** hematopoietic or lymphoid **neoplasms**. However, a specific histology may be covered in more than one module.
Note 3: The modules are **not hierarchical**, but the rules within each module are in **hierarchical** order. Apply the rules **within each module** in order. **Stop** at the first rule that applies
Note 4: Apply rules in Module 1 first. Then go to the **first module** that **applies** to the case you are abstracting. If the situation in your case is not covered in that module **continue** on **as directed** after the last rule in the modules.

Rule	Histology	Other	Code	Notes / Examples
Module 1: General Instructions PH1-PH3 **All hematopoietic and lymphoid neoplasms 9590/3-9992/3**				
PH1	All		Code primary site using • Scans • Medical record documentation • Pathology report • Hematopoietic DB	***Note***: For hematopoietic neoplasms the pathology report is not the automatic default standard for determining the primary site. The standard for determining primary site differs depending upon the specific histology.
PH2	All		Code from **definitive diagnostic method(s)** (See Hematopoietic DB). Definitive diagnostic method can be • Clinical diagnosis • Genetic test • Immunophenotyping • Cytology • Pathology o Final diagnosis o Comment on final diagnosis o Addenda to final diagnosis o CAP protocol	
PH3	When tests or reports defined as definitive diagnosis are not available		Code primary site and histology from medical practitioner's statement on • Medical record • Death certificate	**Go to the appropriate Module 2-8.** **When modules 2-8 do not apply to the case being abstracted, go to Module 9.**
Module 2: Plasma Cell Neoplasms PH4-PH8 **Solitary plasmacytoma of bone 9731/3** **Plasma cell myeloma/multiple myeloma 9732/3**				

Rule	Histology	Other	Code	Notes / Examples
Extraosseous plasmacytoma 9734/3				
PH4	Any of the following occur in a site other than bone • Plasmacytoma • Extraosseous (extramedullary) plasmacytoma • Solitary plasmacytoma • Multiple plasmacytomas • Multiple extraosseous (extramedullary) plasmacytomas		1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ) 2. Histology **extramedullary plasmacytoma** (9734/3)	***Note 1***: Extramedullary and extraosseous mean not occurring in bone ***Note 2***: 80% of extramedullary plasmacytomas occur in the upper respiratory tract (oropharynx, nasopharynx, sinuses, and larynx) although they may occur in numerous other sites including the GI tract, lymph nodes, bladder, CNS, breast, thyroid, testis, parotid, and skin. ***Note 3***: Do **not** code to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424). ***Example 1***: Pathology reports a solitary plasmacytoma wrapped around L4 vertebrae, no invasion of vertebrae. Code the primary site as soft tissue (C496) and the histology 9734/3. ***Example 2***: Scan shows two plasmacytomas in the nasopharyngeal wall. Biopsy confirms plasmacytoma. Code the primary site nasopharynx (C119) and the histology 9734/3.
PH5	Any of the following occur in bone • Plasma cell neoplasm • Solitary plasmacytoma • Solitary plasmacytoma of bone • Solitary medullary plasmacytoma • Multiple plasmacytomas • Multiple plasmacytomas of bone • Multiple medullary		1. Primary site to the **specific bone (C400-C419)** 2. Histology **solitary plasmacytoma of bone (9731/3)**	***Note 1***: The most common sites are bones with active bone marrow hematopoiesis; in order of frequency these include vertebrae, ribs, skull, pelvis, femur, clavicle, and scapula. ***Note 2***: Do **not** code primary site to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424)

Rule	Histology	Other	Code	Notes / Examples
	plasmacytomas of bone • Multiple medullary plasmacytomas			
PH6		Only information is documentation that patient had a plasmacytoma or solitary plasmacytoma	1. Primary site **unknown (C809)** 2. Histology solitary **plasmacytoma of bone (9731/3)**	***Example***: Death certificate only case with underlying cause of death listed as plasmacytoma.
PH7		Clinical diagnosis of plasma cell myeloma/multiple myeloma AND Results of bone marrow biopsy unknown or unavailable	1. Primary site **bone marrow (C421)** 2. Histology plasma cell myeloma/**multiple myeloma (9732/3)**	***Example***: Death-certificate-only case with underlying cause of death listed as multiple myeloma. ***Note***: A clinical diagnosis of multiple myeloma may be based on amyloidosis with associated renal impairment, anemia, and/or hypercalcemia supported by radiologic evidence of multiple lytic bone lesions.
PH8		Diagnosis is Smoldering myeloma Indolent myeloma Evolving myeloma Plasma cell myeloma Multiple myeloma	1. Primary site **bone marrow (C421)** 2. Histology plasma cell myeloma/**multiple myeloma (9732/3)**	***Note*** 1: When the proportion of plasma cells in the bone marrow is 10% or greater, the diagnosis is multiple myeloma. ***Note*** 2: A medical record may have multiple bone marrow biopsies. If any one of the biopsies is positive for multiple myeloma, code the histology to multiple myeloma and the primary site to bone marrow.(C421) ***Example***: Bone marrow Biopsies: Biopsy 1: Negative. Biopsy 2: Multiple myeloma with bone marrow showing 18% plasma cells. Code the primary site bone marrow (C421) and the histology 9732/3. **When this module does not apply to the case being abstracted, go to Module 8.**
Module 3: Lymphoma/Leukemia (Specific neoplasms that can manifest as either leukemia or lymphoma) PH9-PH12 **BCCLL/SLL 9823/3** **Blastic plasmacytoid dendritic cell neoplasm, NOS 9727/3** **Burkitt cell leukemia 9826/3** **Burkitt lymphoma, NOS 9687/3** **Precursor B-cell lymphoblastic leukemia/lymphoma 9836/3** **Precursor B-cell lymphoblastic lymphoma, NOS 9728/3** **Precursor T-cell lymphoblastic lymphoma, NOS 9729/3** **Small B lymphocytic lymphoma 9670/3** **T lymphoblastic leukemia/lymphoma 9670/3**				

Rule	Histology	Other	Code	Notes / Examples
Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma ***Note 2***: Commonly lymphoma originates in lymph node region(s), tissue, or organ(s) although it will metastasize to the bone marrow when the disease is stage IV or disseminated ***Note 3:*** Commonly leukemia originates in the bone marrow.				
PH9		• Diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (BCCLL/SLL) **AND** • There is peripheral blood involvement (bone marrow may also be involved)	1. Primary site **bone marrow (C421)** 2. Histology **B cell chronic lymphocytic leukemia/small lymphocytic lymphoma (BCCLL/SLL) (9823/3)**	***Note 1***: Peripheral blood involvement requires repeated CBCs with absolute lymphocyte count >5000 on repeated measures or flow cytometry that documents a clonal B-cell population in the bone marrow. ***Note 2:*** Leukemic BCCLL will always have peripheral blood involvement. The bone marrow may or may not be involved. In later stages of the disease there may be involvement of lymph nodes, liver and spleen. ***Note 3***: Do not change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.
PH10	B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma	• Diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **AND** • Cannot verify that disease originated in bone marrow	1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ) 2. Histology **small B lymphocytic lymphoma** (9670/3)	***Note 1***: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. ***Note 2***: See Appendix C for help in identifying lymph node regions and codes. ***Note 3***: In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow. ***Note 4***: Small lymphocytic lymphoma is characterized by negative peripheral blood involvement (an absolute lymphocyte count <=5000 on repeated CBCs).
PH11		Diagnosis is • Burkitt lymphoma/leukemia **OR** • Precursor cell lymphoblastic lymphoma/leukemia **OR** • Precursor B-cell	1. Primary site **bone marrow (C421)** 2. Histology (one of the following) • Burkitt cell leukemia (9826/3) • Precursor cell lymphoblastic leukemia, NOS (9835/3) • Precursor B-cell lymphoblastic leukemia (9836/3) • T lymphoblastic leukemia/lymphoma (9837/3)	**Note 1**: Leukemia most commonly originates in the bone marrow. When only the bone marrow is involved, code as leukemia. **Note 2**: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule	Histology	Other	Code	Notes / Examples
		lymphoblastic leukemia/lymphoma **OR** • T-cell lymphoblastic leukemia/lymphoma **AND** • **Only** involvement is bone marrow		
PH12		Diagnosis is • Burkitt lymphoma/leukemia **OR** • Precursor cell lymphoblastic lymphoma/leukemia **OR** • Precursor B-cell 1ymphoblastic leukemia/lymphoma **OR** • Precursor T-cell lymphoblastic leukemia/lymphoma **AND** • Involvement of lymph node region(s), tissue or organ(s)	1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ) 2. Histology (one of the following) • Burkitt lymphoma, NOS (9687/3) • Blastic plasmacytoid dendritic cell neoplasm (previously called precursor cell lymphoblastic lymphoma, NOS) (9727/3) • Precursor B-cell lymphoblastic lymphoma, NOS (9728/3) • Precursor T-cell lymphoblastic lymphoma, NOS (9729/3)	***Note 1***: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. ***Note 2***: See Appendix C for help in identifying lymph node regions, and codes. ***Note 3***: In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow. **When this module does not apply to the case being abstracted, go to Module 8.**
Module 4: Preleukemia, Smoldering Leukemia, and Myelodysplastic Syndrome 9989/3 PH13				
PH13		Diagnosis is • Preleukemia **OR** • Smoldering leukemia **OR** • Myelodysplastic syndrome	1. Primary site **bone marrow (C421)** 2. Histology **myelodysplastic syndrome (9989/3)**	**When this module does not apply to the case being abstracted, go to Module 8.**
Module 5: Myeloid Neoplasms PH14-PH15 **Acute myeloid leukemia, NOS 9861/3** **Myeloid sarcoma 9930/3**				
PH14		• Diagnosis is myeloid neoplasm or acute	1. Primary site **bone marrow (C421)** 2. Histology **acute myeloid leukemia, NOS (9861/3)**	***Note***: Do **not** change primary site code because the spleen is involved with infiltrate.

Rule	Histology	Other	Code	Notes / Examples
		myeloid leukemia, NOS **AND** • Involvement is limited to bone marrow		The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.
PH15		• Diagnosis myeloid neoplasm or myeloid sarcoma **AND** • Neoplasm originates in a site **other than bone marrow**	1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ 2. Histology **myeloid sarcoma (9930/3)**	***Note 1***: Most common sites are skin, lymph node(s), GI tract, bone, soft tissue, and testis. This neoplasm however, can occur in almost every site of the body other than bone. It does **not** originate in bone marrow. ***Note 2***: See Appendix C for help in identifying lymph node names, chains, and codes. **For rules on coding primary site for lymphomas go to Modules 1 and 7.** **When this module does not apply to the case being abstracted, go to Module 8.**
Module 6: Coding Primary Site for Specified Lymphomas PH16-PH24 **Composite lymphoma 9596/3** **Diffuse large B-cell lymphoma 9680/3** **Follicle cell lymphoma 9597/3** **Follicular lymphoma 9690/3** **Follicular lymphoma, grade 1 9695/3** **Follicular lymphoma, grade 2 9691/3** **Follicular lymphoma, grade 3A, 3B 9698/3** **Lymphoplasmacytic lymphoma 9671/3** **Waldenstrom macroglobulinemia 9761/3**				
PH16	• Diffuse large B-cell lymphoma **(DLBCL)** (9680/3) **AND** • **Follicular** lymphoma (9690/3)	DLBCL present in lymph node region(s), tissue, or organ.	1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ) 2. Histology **DLBCL (9680/3)**	***Note 1***: The original pathology may identify only DLBCL although both DLBCL and follicular lymphoma are present. The DLBCL is much more aggressive than the follicular lymphoma and often masks the follicular lymphoma during the initial work-up. Because it is more aggressive, the DLBCL will respond more rapidly to treatment so the post-treatment biopsies may show a combination of DLBCL and follicular lymphoma or the post-treatment biopsy may be positive for only follicular lymphoma. The follicular lymphoma was present from the beginning but was hidden. Do **not** change the histology; it should remain 9680/3 ***Note 2***: Do **not** simply code the site of a

Rule	Histology	Other	Code	Notes / Examples
				biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. ***Note 3***: See Appendix C for help in identifying lymph node names, chains, regions, and codes. ***Note 4***: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they may metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.
PH17	• Diffuse follicular lymphoma **OR** • Follicular lymphoma, diffuse		1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ) 2. Histology **follicular (see examples)**	***Example 1***: Diffuse follicular lymphoma, grade 1. Code follicular lymphoma, grade 1 (9695/3) ***Example 2***: Follicular lymphoma, diffuse, grade 2. Code follicular lymphoma grade 2 (9691/3). ***Example 3***: Grade 3 follicular lymphoma, diffuse. Code follicular lymphoma, grade 3 (9698/3). ***Example 4***: Follicular lymphoma, diffuse. Code follicular lymphoma, NOS (9690/3).
PH18	• **Follicle cell** lymphoma (9597/3) **OR** • **B-cell** lymphoma, **follicle type**	Involvement is limited to • **skin OR** • **skin** and **regional lymph nodes**	1. Primary site to **skin (C44_)** 2. Histology **follicle cell lymphoma (9597/3)**	***Note***: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code follicle cell lymphoma and do not code skin as the primary site. Dissemination to other sites or distant lymph nodes is uncommon and would occur late in the stage of the disease.
PH19	• **Large** B-cell lymphoma **OR** • **B-cell** lymphoma, **large cell type**	Involvement is limited to • **skin OR** • **skin** and **regional lymph nodes**	1. Primary site to **skin (C44_)** 2. Histology **B-cell lymphoma, NOS (9680/3)**	***Note***: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.
PH20	**B-cell** lymphoma, **NOS**	Involvement is limited to • **skin OR** • **skin** and **regional lymph nodes**	1. Primary site to **skin (C44_)** 2. Histology **B-cell lymphoma, NOS (9680/3)**	***Note***: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.
PH21	Both Hodgkin and non-Hodgkin in same lymph node		1. Primary site to **the site of origin** (lymph node region(s), tissue, or organ) 2. Histology **composite lymphoma (9596/3)**	***Note 1***: Use the composite lymphoma code when • Both NHL and HL are present in one

Rule	Histology	Other	Code	Notes / Examples
	region(s), tissue, or organ			lymph node or multiple lymph nodes in one lymph node region. • Both NHL and HL are present in multiple lymph nodes in one lymph node region or several lymph node regions as defined by ICD-O-3. i.e.: NHL and HL present in superior hilum and superior rectal lymph nodes. • Assume all lymph nodes are involved with both NHL and HL even when only one lymph node is biopsied. ***Note 2***: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. ***Note 3***: See Appendix C for help in identifying lymph node names, chains, and codes. ***Note 4***: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If lymph node region(s), tissue, or organ(s) are involved at the time of diagnosis, code as a lymphoma ***Note 5***: Do **not** use the composite lymphoma code 9596/3 when • NHL is present in one node and HL in another node within the same chain i.e.: NHL in one cervical lymph node and HL in another cervical lymph node. • NHL is present in one lymph node region and HL is present in another lymph node region i.e.: NHL in cervical lymph node(s) and HL in inguinal lymph node(s) • NHL in liver and HL in intra-thoracic lymph nodes
PH22	Two or more non-Hodgkin lymphomas present in same lymph node region(s), tissue, or organ		1. Primary site to the **site of origin** (lymph node region(s), tissue, or organ) 2. Histology **NHL with numerically highest ICD-O-3 code**	***Note 1***: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. ***Note 2***: See Appendix C for help in identifying lymph node names, chains,

Rule	Histology	Other	Code	Notes / Examples
				regions, and codes. ***Note 3***: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If lymph node region(s), tissue, or organ(s) are involved at the time of diagnosis, code as a lymphoma. ***Note 4***: This rule does not apply when NHL is present in different sites. Examples are • Thymic extranodal marginal-zone B-cell lymphoma is present in the thymus and diffuse large B-cell lymphoma in the hilar lymph nodes. • B-cell lymphoma is present in the intrathoracic lymph nodes and peripheral T-cell NHL in the liver.
PH23	Waldenstrom macroglobulinemia	• Lymphoplasmacytic lymphoma in the bone marrow **AND** • IgM monoclonal gammopathy in the blood	1. Primary site to **blood (C420)** 2. Histology **Waldenstrom macroglobulinemia (9761/3)**	
PH24	• Waldenstrom macroglobulin-emia **OR** • Lymphoplasma-cytic lymphoma **AND** Waldenstrom macroglobulin-emia	Bone marrow, lymph nodes **AND/OR** tissue are involved	1. Primary site to involved **bone marrow, lymph nodes, or tissue** 2. Histology **lymphoplasmacytic lymphoma (9671/3)**	**For additional rules on coding primary site for lymphomas go to Modules 1 and 7. When this module does not apply to the case being abstracted, go to Module 8**
Module 7: Primary Site Rules for Lymphomas Only 9590/3-9729/3 PH25-PH37				
PH25		Involvement of • Only one lymph node **OR** • One lymph node chain	Primary site to **specific lymph node region**	
PH26		Site of lymphoma described only as a mediastinal mass	Primary site to **mediastinal lymph nodes (C771)**	
PH27		Site of lymphoma described only as a retroperitoneal mass **OR**	Primary site to **intra-abdominal lymph nodes (C772)**	

Rule	Histology	Other	Code	Notes / Examples
		as a mesenteric mass		
PH28		Site of lymphoma described only as an inguinal mass	Primary site to **inguinal lymph nodes (C774)**	
PH29		Involvement of multiple lymph node chains within the same region	Primary site to **specific lymph node region as defined by ICD-O-3**	***Example 1***: Code intra-abdominal lymph nodes (C772) when there is involvement of hepatic (C772) and para-aortic lymph node chains (C772). ***Example 2***: Code lymph nodes of head, face and neck (C770) when there is involvement of cervical (C770) and mandibular (C770) lymph node chains. ***Example 3***: Code mediastinal lymph nodes (C771) when bilateral mediastinal lymph nodes are involved.
PH30		• Involvement of multiple lymph node regions (as defined by ICD-O-3) **AND** • Lymph node region where lymphoma originated cannot be identified	Primary site to multiple lymph node regions, NOS (C778)	***Note 1***: Do not simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma. ***Note 2***: See Appendix C for help in identifying lymph node names, chains, regions, and codes. ***Example 1***: Cervical and intrathoracic lymph nodes involved with B-cell lymphoma. Code the primary site to lymph nodes of multiple regions (C778). ***Example 2***: CT scans showed involvement of the cervical lymph nodes (C770) and the mediastinal lymph nodes (C771). No additional involvement was identified during the work-up. Biopsy of a cervical lymph node confirmed lymphoma. Code the primary site to lymph nodes of multiple regions (C778).
PH31		• Lymph nodes are involved **AND** • No particular primary site/lymph node region is identified	Primary site to **lymph nodes, NOS (C779)**	
PH32		Lymphoma present only in bone marrow	Primary site to **bone marrow (C421)**	***Note***: All available physical exams, scans, and other work-up must be negative for lymph node, tissue, or organ involvement.
PH33		Lymphoma present only in	Primary site to the **organ**	***Note***: Includes lymphomas that are primary in

Rule	Histology	Other	Code	Notes / Examples
		one organ		the spleen. Although these lymphomas are rare, if the physician states that spleen is the organ of origin, code the primary site spleen (C422). ***Example***: Pathology from stomach resection shows lymphoma. No other pathologic or clinical disease identified. Code the primary site to stomach, NOS (C169)
PH34		There is proof of extension from regional lymph nodes into an organ.	Primary site to lymph node region as defined by ICD-O-3	***Example***: Patient presents with abdominal adenopathy. Surgical exploration documents direct invasion of the stomach from the regional lymph nodes. Code abdominal lymph nodes (C772).
PH35		Lymphoma present in: • An organ **AND** • That organ's **regional** lymph nodes ONLY	Primary site to the **organ**	***Note***: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes. ***Example 1***: Lymphoma is present in the kidney and peri-renal lymph nodes. Code the primary site to kidney (C649). ***Example 2***: Lymphoma is present in the stomach and the gastric lymph nodes. Code the primary site to stomach, NOS (C169). ***Example 3***: Lymphoma is present in the spleen and the splenic lymph nodes. Code the primary site spleen (C422).
PH36		Lymphoma present in • An organ **AND** • Lymph nodes that are **not** regional for that organ **AND** • Site of origin cannot be determined (even after consulting physician)	Primary site to **lymph nodes, NOS (C779)**	***Note 1***: Lymphoma can spread from organs to regional lymph nodes, but does not spread from the organ directly to distant lymph nodes ***Example***: The patient has positive mediastinal C771 and cervical (C770 lymph nodes and involvement of the stomach (C169). No further information is available. Code to lymph node, NOS (C779) ***Note 2***: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes. ***Note 3***: See Appendix C for help in identifying lymph node names, chains, regions, and codes.
PH37		• No evidence of lymphoma in lymph nodes **AND** • Physician documents	Primary site to **unknown primary (C809)**	**See ICD-O-3 Rule D**

Rule	Histology	Other	Code	Notes / Examples
		that he/she suspects that the lymphoma originated in an organ.		**When this module does not apply to the case being abstracted, go to Module 8.**
Module 8: Histology Rules Only: All hematopoietic and lymphoid neoplasms 9590/3-9992/3 PH38 - PH39				
PH38		• **One non-specific** histology **AND** • **Two or more specific** histologies **AND** • The hematopoietic DB multiple primaries calculator documents the specific histologies and NOS are the **same primary AND** • No further information is available	Histology to non-specific histology	***Note 1***: Use Appendix E: Histology "NOS" Tables to identify the NOS histologies. ***Note 2***: Use the Hematopoietic DB multiple primaries calculator to confirm that the NOS and specific histologies are the same primary. ***Example***: The diagnosis myeloproliferative disorder, NOS (9960/3), polycythemia vera (9950/3), essential thrombocythemia (9962/3). The Hematopoietic DB multiple primaries calculator shows myeloproliferative disorder and polycythemia vera are the same primary. The multiple primaries calculator also shows myeloproliferative disorder and essential thrombocythemia are the same primary. Follow-back produces no additional information. Code the histology myeloproliferative disorder, NOS (9960/3).
PH39		• **One non-specific (NOS)** histology **AND** • **One specific** histology **AND** • The Hematopoietic DB multiple primaries calculator documents the specific histology and NOS are the **same primary**	Histology to the **specific** histology	***Note 1***: Use Appendix E: Histology "NOS" Tables to identify the NOS histologies. ***Note 2***: Use the Hematopoietic DB multiple primaries calculator to confirm that the NOS and specific histology are the same primary. **When this module does not apply to the case being abstracted, go to Module 9**
Module 9: Default Rules: All hematopoietic and lymphoid neoplasms 9590/3-9992/3 PH40-PH41				
PH40		When rules PH1-PH39 do **not** apply	Primary site using **Hematopoietic DB** Histology using **Hematopoietic DB**	
PH41		Cannot determine histology code by using the Hematopoietic DB	Histology to the **numerically higher ICD-O-3 code.**	**This is the end of the rules for coding primary site and histology.**

Grade of Tumor Rules Matrix Format

Note 1: Use the Grade of Tumor Rules (G1-G11) to assign the correct code in the grade field.
Note 2: Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade.

Rule	Statement in Medical Record	Code	Comments / Notes
G1	Myeloproliferative neoplasms, myeloproliferative/myelodysplastic syndromes, myelodysplastic syndrome, histiocytic and dendritic cell neoplasms 9740/3 9741/3 9742/3 9751/3 9755/3 9757/3 9758/3 9759/3 9801/3 9805/3 9806/3 9807/3 9808/3 9809/3 9875/3 9876/3 9945/3 9946/3 9950/3 9961/3 9962/3 9963/3 9964/3 9975/3 9980/3 9982/3 9982/3 9983/3 9985/3	Cell type not determined, not stated, not applicable (**code 9**)	***Note 1:*** These neoplasms do not have a specific codable phenotype ***Note 2***: See Tables B1, B3, B4, and B11 in Appendix B for neoplasm terms and codes.

Rule	Statement in Medical Record	Code	Comments / Notes
	9986/3 9989/3 9991/3 9992/3		
G2	Use statements from **any part** of medical record including but not limited to • Pathology report **OR** • History and physical **OR** • Consultation **OR** • Final diagnosis **OR** • Face sheet		N/A
G3	All hematopoietic and lymphoid neoplasms	**Only** codes 5, 6, 7, 8, **OR** 9	***Note 1***: Do **not** code descriptions "low grade," intermediate grade," or "high grade" in the Tumor Grade field. These terms refer to the Working Formulation categories of lymphoma diagnosis. ***Note 2***: Do **not** code the descriptions "Grade 1," "Grade 2," or "Grade 3" in the Tumor Grade field. These grades represent histology types of lymphoma rather than differentiation.
G4	T-cell is part of the neoplasm name **OR** Neoplasm is of T-cell origin 9700/3 9701/3 9701/3 9702/3 9705/3 9708/3 9709/3 9714/3 (unless pathologist specifically designates as a B-cell) 9716/3 9717/3 9718/3 9724/3 9725/3 9726/3 9827/3 9831/3 9834/3 9837/3	T-cell (**code 5)**	***Note 1***: Record T-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention T-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name. ***Note 2***: When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.
G5	• T-cell **OR** • T-cell phenotype **OR**	T-cell (**code 5**)	N/A

Rule	Statement in Medical Record	Code	Comments / Notes
	• T-precursor **OR** • Pre-T, gamma-delta T **OR** • Null cell and T-cell		
G6	**B-cell precursor lymphoid neoplasms** and the **mature B-cell neoplasms** 9591/3 9596/3 9597/3 9670/3 9671/3 9673/3 9678/3 9679/3 9680/3 9684/3 9687/3 9688/3 9689/3 9690/3 9691/3 9695/3 9698/3 9699/3 9712/3 9728/3 9731/3 9732/3 9734/3 9737/3 9738/3 9762/3 9811/3 9812/3 9813/3 9814/3 9815/3 9816/3 9817/3 9818/3 9823/3 9833/3	B-cell (**code 6**)	***Note 1***: Record B-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention B-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name. ***Note 2***: When the medical record or pathology report contains one of these terms with a different phenotype (T-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification. ***Note 3***: See Tables B7 and B8 in Appendix B

Rule	Statement in Medical Record	Code	Comments / Notes
	9836/3 9940/3		
G7	• B-cell, B-cell phenotype **OR** • B-precursor **OR** • Pre-B **OR** • Null-cell and B-cell	B-cell (**code 6**)	N/A
G8	• Null cell OR • Non-T non-B OR • Common cell	Null-cell, non-T non-B (**code 7**)	N/A
G9	NK-cell is part of the neoplasm's name 9719/3 9948/3	NK (natural killer) (**code 8**)	***Note 1***: Record NK-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention NK-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name. ***Note 2***: When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, T-cell, or null-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification. ***Note 3***: See Table B9 in Appendix B
G10	• NK cell **OR** • Natural killer cell **OR** • Nasal NK/T-cell lymphoma **OR** • Null-cell and NK cell	NK (natural killer) (**code 8**)	N/A
G11	• No statement describing cell type **OR** • Cell type is described as combined T **AND** B cell **OR** • Cell type is described as **combined** B **AND** NK cell	Cell type not determined, not stated, not applicable (**code 9**)	***Note***: There is a new site-specific factor to collect combination cell types for hematopoietic or lymphatic neoplasms in the Collaborative Stage Data Collection System, Version 2

Multiple Primary Rules Text Format

Note 1: Use Multiple Primary Rules M1 through M12 **before** using the Hematopoietic DB.
Note 2: The registrar must recognize that during the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology is identified. These diagnoses are not multiple primaries; they represent steps in the diagnostic work-up. See rules M7 - M12.

Rule M1 Abstract as a single primary when **minimal information** is available (such as a death certificate only (DCO) case or a pathology-report-only case). *

Rule M2 Abstract as a single primary when there is a **single histology.***
Example 1: The diagnosis is multiple myeloma (9732/3). Abstract as a single primary.
Example 2: Multiple extraosseous plasmacytomas (9734/3) are present in the oropharynx. Abstract as a single primary.
Example 3: A single histology diagnosed by the definitive diagnostic method as defined in the Hematopoietic DB; **for example**. The patient had several provisional diagnoses but the definitive diagnostic method identifies a single histology. Abstract as a single primary.

Rule M3 Abstract as a single primary when **two or more types of non-Hodgkin lymphoma** are present in the same anatomic location(s), such as one lymph node, one organ, or one tissue.*
Example: Biopsy of cervical lymph node shows **two** different non-Hodgkin lymphomas. Abstract as a single primary.
Note 1: Do **NOT** use this rule for cutaneous lymphomas.
Note 2: When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for information on lymph node codes and regions.
Note 3: When the disease is in a more advanced stage, both non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3 or in an organ and that organ's regional lymph nodes or in multiple organs.
- Although both non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination non-Hodgkin lymphoma, assume that all of the nodes, tissue, and/or organs are involved with the combination of non-Hodgkin lymphomas.

Note 4: Do **not** query the Hematopoietic DB in this situation.

Rule M4 Abstract as a single primary when both **Hodgkin and non-Hodgkin** lymphoma are present in the **same** anatomic **location(s)**. Hodgkin and non-Hodgkin may be present in one lymph node, one organ, or one tissue.*

Example: Biopsy of cervical lymph node shows Hodgkin and non-Hodgkin lymphomas. Abstract as a single primary.

Note 1: When the disease is in an early stage, the involved lymph node(s) will be in the same region as defined by ICD-O-3 codes. See Appendix C for lymph node codes and regions.

Note 2: When the disease is in a more advanced stage, both Hodgkin and non-Hodgkin lymphomas may be present in multiple lymph node regions as defined by ICD-O-3 codes or in an organ and that organ's regional lymph nodes or in multiple organs.

- Although both Hodgkin and non-Hodgkin lymphomas must be present in each of the involved sites in order to abstract as a single primary, it is not required that all involved organs be biopsied. If the physician biopsies one of the involved sites and diagnoses the combination Hodgkin and non-Hodgkin lymphomas, assume that all of the nodes, tissue, and/or organs are involved with the combination of Hodgkin and non-Hodgkin lymphomas.

Note 3: Do **not** query the Hematopoietic DB in this situation.

Rule M5 Abstract as multiple primaries when any of the following situations are met **

- Hodgkin lymphoma in **one node** and non-Hodgkin lymphoma in a **different node**

  Note: The involved nodes may be in the same lymph node region as defined by ICD-O-3 or in different lymph node regions as defined by ICD-O-3. See Appendix C.

- Hodgkin lymphoma in **one organ** and non-Hodgkin lymphoma in a **different organ**

- Hodgkin lymphoma in **tissue** and non-Hodgkin lymphoma in **different tissue**

  Example 1: Patient is diagnosed with Hodgkin lymphoma in the cervical lymph nodes and also with non-Hodgkin lymphoma in the inguinal lymph nodes. Abstract as multiple primaries.

  Example 2: Hodgkin lymphoma in thymus and non-Hodgkin lymphoma in the tonsil. Abstract as multiple primaries.

  Example 3: Hodgkin lymphoma in the brain and non-Hodgkin lymphoma in the mediastinal lymph nodes. Abstract as multiple primaries.

Rule M6 Abstract as a single primary when a **more specific** histology is diagnosed after an **NOS** when the Hematopoietic DB Multiple Primaries Calculator confirms that the NOS and the more specific histology are the same primary.

Note 1: There are no time restrictions on these diagnoses; the interval between the NOS and the more specific histology does not affect this rule stating that the two neoplasms are a single primary.

Note 2: The Hematopoietic DB will identify these histology ies as a single primary.

Rule M7 Abstract as a single primary when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days **AND ***

- There is documentation of one positive bone marrow biopsy

Note: When these diagnoses happen within 21 days, it is highly possible that one diagnosis was provisional and the bone marrow identified the correct diagnosis.

Rule M8 Abstract as multiple primaries when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days **AND ****

- There is documentation of two bone marrow examinations, one confirming the chronic neoplasm and another confirming the acute neoplasm

Rule M9 Abstract as a single primary when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days **AND ***

- There is no available documentation on bone marrow biopsy

Note 1: The two diagnoses are likely the result of an ongoing diagnostic work-up. The later diagnosis is usually based on all of the test results.

Note 2: This rule applies if both neoplasms are diagnosed simultaneously (at the same time).

Rule M10 Abstract as multiple primaries when a neoplasm is **originally diagnosed** in a **chronic** (less aggressive) phase AND **second diagnosis** of a blast or acute phase **more than 21 days** after the chronic diagnosis. **

Note 1: **This is a change from previous rules**. Use the Hematopoietic DB to determine multiple primaries when a transformation from the chronic to a blast or acute phase occurs.

Note 2: Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.

Rule M11 Abstract the acute phase as a single primary when a neoplasm is **originally diagnosed** in the blast or acute phase and **reverts** to a less aggressive/chronic phase and there is **no confirmation** available that the patient has been treated.

Note 1: When these diagnoses happen **within 21 days**, it is highly possible that the first diagnosis of acute disease was a provisional diagnosis.

Note 2: When the subsequent diagnosis occurs more than 21 days after the original diagnosis of acute disease it is important to follow-back to obtain information on treatment or a subsequent bone marrow biopsy that negates the diagnosis of acute disease.

Rule M12 Abstract as multiple primaries when a neoplasm is **originally diagnosed** in the blast or acute phase and **reverts** to a less aggressive/chronic phase **after treatment. ****

Note 1: Only abstract as a multiple primary when the patient has been treated for the acute disease.

Note 2: **This is a change from previous rules.** Use the Hematopoietic DB to determine multiple primaries when a transformation from the blast or acute phase to a chronic phase occurs.

Note 3: Transformations are defined in the Hematopoietic DB for each hematopoietic and lymphoid neoplasm.

Rule M13 Use the Hematopoietic DB to determine the number of primaries for all cases that do **not** meet the criteria of M1-M12.

*Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.
**Prepare two or more abstracts. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes to each case abstracted.

Primary Site and Histology Coding Rules Text Format

Note 1: Use the Primary Site and Histology Rules **before** using the Hematopoietic DB
Note 2: The primary site and histology coding rules are divided into nine modules. Each module covers a group of **related** hematopoietic or lymphoid **neoplasms**. However, a specific histology may be covered in more than one module.
Note 3: The modules are **not hierarchical**, but the **rules** within each module **are** in **hierarchical** order. Apply the rules **within each module** in order. **Stop** at the first rule that applies.
Note 4: Apply rules in Module 1 first. Then go to the **first module** that **applies** to the case you are abstracting. If the situation in your case is not covered in that module **continue** on as **directed** after the last rule in the module.

Module 1: General Instructions PH1-PH3

All hematopoietic and lymphoid neoplasms (9590/3-9992/3)

Rule PH1 Code the primary site using information from scans, documentation in the medical record, the pathology report, and from the Hematopoietic DB.
Note: For hematopoietic neoplasms the pathology report is not the automatic default standard for determining the primary site. The standard for determining primary site differs depending upon the specific histology.

Rule PH2 Code the histology diagnosed by the definitive diagnostic method(s) (see Hematopoietic DB). The definitive diagnostic method can be a clinical diagnosis, genetic test, immunophenotyping, cytology, or pathology. When a pathology report is the definitive diagnostic method, code the histology from the final diagnosis, comment on the final diagnosis, addenda to the final diagnosis, or CAP protocol.

Rule PH3 Code the primary site and histology using the medical practitioner's statement on the medical record or death certificate when none of the tests or reports defined as a definitive diagnostic method is available.

Go to the appropriate Module 2-8.
When modules 2-8 do not apply to the case being abstracted, go to Module 9.

Module 2: Plasma Cell Neoplasms PH4-PH8

Solitary plasmacytoma of bone 9731/3
Plasma cell myeloma/multiple myeloma 9732/3
Extraosseous plasmacytoma 9734/3

Rule PH4 Code the primary site to the site of origin, (lymph node region(s), tissue, or organ) and code the histology extramedullary plasmacytoma (9734/3) when any of the following occur in a site other than bone

- **Plasmacytoma**
- **Extraosseous (extramedullary) plasmacytoma**
- **Solitary plasmacytoma**
- **Multiple plasmacytomas**
- **Multiple extraosseous (extramedullary) plasmacytomas**

Note 1: Extramedullary and extraosseous mean "not occurring in bone."

Note 2: 80% of extramedullary plasmacytomas occur in the upper respiratory tract (oropharynx, nasopharynx, sinuses, and larynx) although they may occur in numerous other sites including the GI tract, lymph nodes, bladder, CNS, breast, thyroid, testis, parotid, and skin.

Note 3: Do **not** code to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

Example 1: Pathology reports a solitary plasmacytoma wrapped around L4 vertebrae, no invasion of vertebrae. Code the primary site as soft tissue (C496) and the histology 9734/3.

Example 2: Scan shows two plasmacytomas in the nasopharyngeal wall. Biopsy confirms plasmacytoma. Code the primary site nasopharynx (C119) and the histology 9734/3.

Rule PH5 Code the primary site to the **specific bone (C400-C419)** where the plasmacytoma originated and code the histology solitary plasmacytoma of bone (9731/3) when the diagnosis is

- **Plasma cell neoplasm**
- **Solitary plasmacytoma**
- **Solitary plasmacytoma of bone**
- **Solitary medullary plasmacytoma**
- **Multiple plasmacytomas**
- **Multiple plasmacytomas of bone**
- **Multiple medullary plasmacytomas**

Note 1: The most common sites are bones with active bone marrow hematopoiesis; in order of frequency these include vertebrae, ribs, skull, pelvis, femur, clavicle, and scapula.

Note 2: Do **not** code primary site to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

Rule PH6 Code the primary site **unknown (C809)** and histology solitary plasmacytoma of bone (9731/3 when the only information is that the patient had a **plasmacytoma** or a **solitary plasmacytoma**.

Example: Death-certificate-only case with underlying cause of death listed as plasmacytoma.

Rule PH7 Code the primary site **bone marrow (C421)** and the histology plasma cell myeloma/**multiple myeloma** (9732/3) when the **clinical** diagnosis is plasma cell myeloma/multiple myeloma and there is no documentation of bone marrow biopsy or the results of the bone marrow biopsy are unknown or unavailable.

Example: Death-certificate-only case with underlying cause of death listed as multiple myeloma.

Note: A clinical diagnosis of multiple myeloma may be based on amyloidosis with associated renal impairment, anemia, and/or hypercalcemia supported by radiologic evidence of multiple lytic bone lesions.

Rule PH8 Code the primary **site bone marrow (C421)** and the histology plasma cell myeloma/**multiple myeloma** (9732/3) when the diagnosis is **smoldering myeloma, indolent myeloma, evolving myeloma, plasma cell myeloma,** or **multiple myeloma.**

Note 1: When the proportion of plasma cells in the bone marrow is 10% or greater, the diagnosis is multiple myeloma.

Note 2: A medical record may have multiple bone marrow biopsies. If any one of the biopsies is positive for multiple myeloma, code the histology to multiple myeloma (9732/3) and the primary site to bone marrow (C421).

Example: Bone marrow biopsies: Biopsy 1: Negative. Biopsy 2: Multiple myeloma with bone marrow showing 18% plasma cells. Code the primary site bone marrow (C421) and the histology 9732/3.

When this module does not apply to the case being abstracted, go to Module 8.

Module 3: Lymphoma/Leukemia (Specific neoplasms that can manifest as either leukemia or lymphoma) PH9-PH12

Blastic plasmacytoid dendritic cell neoplasm, NOS 9727/3
Burkitt cell leukemia 9826/3
Burkitt lymphoma, NOS 9687/3
Precursor B-cell lymphoblastic leukemia/lymphoma 9836/3
Precursor B-cell lymphoblastic lymphoma, NOS 9728/3
Precursor T-cell lymphoblastic lymphoma, NOS 9729/3
Small B lymphocytic lymphoma 9670/3
T lymphoblastic leukemia/lymphoma 9670/3

Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma
Note 2: Commonly lymphoma originates in lymph node region(s), tissue, or organ(s) although it will metastasize to the bone marrow when the disease is stage IV or disseminated
Note 3: Commonly leukemia originates in the bone marrow

Rule PH9 Code the primary site **bone marrow (C421)** and code the histology B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **(BCCLL/CLL) (9823/3)** when the diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (**BCCLL/SLL**) AND **peripheral blood** is **involved** (the **bone marrow** may also be involved).

Note 1: Peripheral blood involvement requires repeated CBCs with absolute lymphocyte count >5000 on repeated measures or flow cytometry that documents a clonal B-cell population in the bone marrow.

Note 2: Leukemic BCCLL will always have peripheral blood involvement. The bone marrow may or may not be involved. In later stages of the disease there may be involvement of lymph nodes, liver and spleen.

Note 3: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH10 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and code the histology **small B lymphocytic lymphoma (9670/3)** when the diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma **AND** you **cannot verify** that the disease originated in the bone marrow.

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma.

Note 2: See Appendix C for help in identifying lymph node regions and codes.

Note 3: In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow.

Note 4: Small lymphocytic lymphoma is characterized by negative peripheral blood involvement (an absolute lymphocyte count <=5000 on repeated CBCs).

Rule PH11 Code the primary site **bone marrow (C421)** and the respective histology from the list below when the diagnosis is Burkitt **lymphoma/leukemia**, precursor cell lymphoblastic lymphoma/leukemia, precursor B-cell 1ymphoblastic leukemia/lymphoma, or precursor T-cell lymphoblastic leukemia/lymphoma AND the **only** involvement is **bone marrow**.

- Burkitt cell leukemia (9826/3)
- Precursor cell lymphoblastic leukemia, NOS (9835/3)
- Precursor B-cell lymphoblastic leukemia/lymphoma (9836/3)
- T lymphoblastic leukemia/lymphoma (9837/3)

Note 1: Leukemia most commonly originates in the bone marrow. When only the bone marrow is involved, code as leukemia.

Note 2: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH12 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology to the respective histology from the list below when the diagnosis is Burkitt **lymphoma/leukemia**, precursor cell lymphoblastic lymphoma/leukemia, precursor B-cell lymphoblastic leukemia/lymphoma, or precursor T-cell lymphoblastic leukemia/lymphoma **AND** there is involvement of lymph node region(s), tissue, or organ(s).

- Burkitt lymphoma, NOS (9687/3)
- Blastic plasmacytoid dendritic cell neoplasm (previously called precursor cell lymphoblastic lymphoma, NOS) (9727/3)
- Precursor B-cell lymphoblastic lymphoma, NOS (9728/3)
- Precursor T-cell lymphoblastic lymphoma, NOS (9729/3)

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma.

Note 2: See Appendix C for help in identifying lymph node regions, and codes.

Note 3: In early stages of this lymphoma (Stage I, Stage II), only lymph nodes are involved. In later stages (Stage III, Stage IV) there may be involvement of the liver, spleen and/or bone marrow.

When this module does not apply to the case being abstracted, go to Module 8.

Module 4: Preleukemia, Smoldering leukemia and Myelodysplastic syndrome 9989/3
PH13

Rule PH13 Code the primary site **bone marrow (C421)** and the histology **myelodysplastic syndrome (9989/3)** when the diagnosis is **preleukemia, smoldering leukemia, or myelodysplastic syndrome.**

When this module does not apply to the case being abstracted, go to Module 8.

Module 5: Myeloid Neoplasms PH14-PH15

Acute myeloid leukemia, NOS 9861/3
Myeloid sarcoma 9930/3

Rule PH14 Code the primary site **bone marrow (C421)** and code the histology **9861/3** when the diagnosis is **myeloid neoplasm** or **acute myeloid leukemia**, **NOS AND** the involvement is **limited to bone marrow.**

Note: Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.

Rule PH15 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ and the histology to **myeloid sarcoma (9930/3)** when the diagnosis is **myeloid neoplasm** or **myeloid sarcoma AND** the neoplasm originates in a site **other than bone marrow.**

Note 1: Most common sites are skin, lymph node(s), GI tract, bone, soft tissue, and testis. This neoplasm however, can occur in almost every site of the body other than bone. Myeloid sarcoma does **not** originate in bone marrow.

Note 2: See Appendix C for help in identifying lymph node names, chains, and codes.

For rules on coding primary site for lymphomas go to Modules 1 and 7.
When this module does not apply to the case being abstracted, go to Module 8.

Module 6: Coding Primary Site and Histology for Specified Lymphoma PH16-PH24

Composite lymphoma 9596/3
Diffuse large B-cell lymphoma 9680/3
Follicle cell lymphoma 9597/3
Follicular lymphoma 9690/3
Follicular lymphoma, grade 1 9695/3
Follicular lymphoma, grade 2 9691/3
Follicular lymphoma, grade 3A, 3B 9698/3
Lymphoplasmacytic lymphoma 9671/3
Waldenstrom macroglobulinemia 9761/3

Rule PH16 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and code the histology diffuse large B-cell lymphoma (DLBCL) (9680/3) when **DLBCL** (9680/3) and **follicular lymphoma** (9690/3) are present **in the same lymph node**(s), **tissue, or organ**

Note 1: The original pathology may identify only DLBCL although both DLBCL and follicular lymphoma are present. The DLBCL is much more aggressive than the follicular lymphoma and often masks the follicular lymphoma during the initial work-up. Because it is more aggressive, the DLBCL will respond more rapidly to treatment so the post-treatment biopsies may show a combination of DLBCL and follicular lymphoma or the post-treatment biopsy may be positive for only follicular lymphoma. The follicular lymphoma was present from the beginning but was hidden. Do not change the histology; it should remain 9680/3

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma.

Note 3: See Appendix C for help in identifying lymph node names, chains, regions, and codes.

Note 4: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.

Rule PH17 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology to follicular when the lymphoma is described as diffuse follicular or follicular, diffuse

Example 1: Diffuse follicular lymphoma, grade 1. Code follicular lymphoma, grade 1 (9695/3)

Example 2: Follicular lymphoma, diffuse, grade 2. Code follicular lymphoma grade 2 (9691/3).

Example 3: Grade 3 follicular lymphoma, diffuse. Code follicular lymphoma, grade 3 (9698/3).

Example 4: Follicular lymphoma, diffuse. Code follicular lymphoma, NOS (9690/3).

Rule PH18 Code the primary site to **skin** (C44_) and the histology to **follicle cell lymphoma** (9597/3) when there is **skin infiltration** with **follicle cell lymphoma** or **B-cell lymphoma, follicle type** and the involvement is limited to **skin**, or limited to **skin** and the **regional lymph nodes**.

Note: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do not code follicle cell lymphoma and do **not** code skin as the primary site. Dissemination to other sites or distant lymph nodes is uncommon and would occur late in the stage of the disease.

Rule PH19 Code the primary site to **skin** (C44_) and the histology to **large B-cell lymphoma** (9680/3) when there is **skin infiltration** with **large B-cell lymphoma** or **B-cell lymphoma**, **large cell type** and the involvement is limited to **skin**, or limited to **skin** and the **regional lymph nodes**.

Note: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.

Rule PH20 Code the primary site to **skin** (C44_) and the histology to **B-cell lymphoma, NOS** (9680/3) when there is **skin infiltration** with **B-cell lymphoma** and the involvement is limited to **skin**, or limited to **skin** and the **regional lymph nodes**.

Note: If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do **not** code skin as the primary site.

Rule PH21 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology **composite lymphoma (9596/3)** when **both** non-Hodgkin lymphoma and Hodgkin lymphoma are **present in the same lymph node** region(s), tissue, or organ

Note 1: Use the composite lymphoma code when

- Both NHL and HL are present in one lymph node or multiple lymph nodes in one lymph node region.
- Both NHL and HL are present in multiple lymph nodes in one lymph node region or several lymph node regions as defined by ICD-O-3. i.e.: NHL and HL present in superior hilum and superior rectal lymph nodes.
    - Assume all lymph nodes are involved with both NHL and HL even when only one lymph node is biopsied.

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Module 1 and 7 for more information on coding primary site for lymphoma.

Note 3: See Appendix C for help in identifying lymph node names, chains, and codes.

Note 4: Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma

Note 5: Do **not** use the composite lymphoma code 9596/3 when:

- NHL is present in one lymph node region and HL is present in another lymph node region i.e.: NHL in cervical lymph node(s) and HL in inguinal lymph node(s)
- NHL in liver and HL in intra-thoracic lymph nodes

Rule PH22 Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology to the **numerically highest ICD-O-3 code** when two or more **non-Hodgkin lymphomas** are present in the same lymph node(s), tissue, or organ.

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma.

Note 2: See Appendix C for help in identifying lymph node names, chains, regions, and codes.

Note 3: Commonly lymphomas originate in lymph node region(s), tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.

Note 4: This rule does not apply when NHL is present in different sites. Examples are:

- Thymic extranodal marginal-zone B-cell lymphoma is present in the thymus and diffuse large B-cell lymphoma in the hilar lymph nodes.
- B-cell lymphoma is present in the intrathoracic lymph nodes and peripheral T-cell NHL in the liver.

Rule PH23 Code the primary site blood (C420) and the histology Waldenstrom macroglobulinemia (9761/3) when there is lymphoplasmacytic lymphoma in the bone marrow and IgM monoclonal gammopathy in the blood.

Rule PH24 Code the primary site to the involved bone marrow, lymph nodes, or lymphoid tissue and the histology lymphoplasmacytic lymphoma (9671/3) when the diagnosis is Waldenstrom macroglobulinemia OR lymphoplasmacytic lymphoma and Waldenstrom macroglobulinemia AND the bone marrow, lymph nodes OR lymphoid tissue are involved.

For additional rules on coding primary site for lymphomas go to Module 1 and 7.
When this module does not apply to the case being abstracted, go to Module 8.

Module 7: Primary Site Rules for Lymphomas Only 9590/3-9729/3
PH25-PH37

Rule PH25 Code the primary site to the specific lymph node region when only **one lymph node or one lymph node region** is involved.

Rule PH26 Code the primary site mediastinal lymph nodes (C771) when the site of lymphoma is described **only** as a **mediastinal mass.**

Rule PH27 Code the primary site intra-abdominal lymph nodes (C772) when the site of lymphoma is described **only** as a **retroperitoneal mass** or as a **mesenteric mass.**

Rule PH28 Code the primary site inguinal lymph nodes (C774) when the site of lymphoma is described only as an inguinal mass.

Rule PH29 Code the primary site to the specific lymph node region when **multiple** lymph node **chains** within the **same region** (as defined by ICD-O-3) are involved.

Example 1: Code intra-abdominal lymph nodes (C772) when there is involvement of hepatic (C772) and para-aortic lymph node chains (C772).
Example 2: Code lymph nodes of head, face and neck (C770) when there is involvement of cervical (C770) and mandibular (C770) lymph node chains.
Example 3: Code mediastinal lymph nodes (C771) when bilateral mediastinal lymph nodes are involved.

Rule PH30 Code the primary site as multiple lymph node regions, NOS when multiple lymph node regions (C778) as defined by ICD-O-3 are involved and it is **not possible to identify the lymph node region where the lymphoma originated**.

Note 1: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma.
Note 2: See Appendix C for help in identifying lymph node names, chains, regions, and codes.
Example 1: Cervical and intrathoracic lymph nodes involved with B-cell lymphoma. Code the primary site to lymph nodes of multiple regions (C778).
Example 2: CT scans showed involvement of the cervical lymph nodes (C770) and the mediastinal lymph nodes (C771). No additional involvement was identified during the work-up. Biopsy of a cervical lymph node confirmed lymphoma. Code the primary site to lymph nodes of multiple regions (C778).

Rule PH31 Code the primary site to lymph nodes, NOS (C779) when lymph node(s) are involved but **no primary site/particular lymph node region** is identified.

Rule PH32 Code the primary site to bone marrow (C421) when lymphoma is **present only in the bone marrow.**

Note: All available physical exams, scans, and other work-up must be negative for lymph node, tissue, or organ involvement.

Rule PH33 Code the primary site to the specific organ when **lymphoma** is present only in an **organ**.

Note: Includes lymphomas that are primary in the spleen. Although these lymphomas are rare, if the physician states that spleen is the organ of origin, code the primary site spleen (C422).
Example: Pathology from stomach resection shows lymphoma. No other pathologic or clinical disease identified. Code the primary site to stomach, NOS (C169).

Rule PH34 Code the primary site to the lymph node region as defined by ICD-O-3 when there **is proof of extension from the regional lymph nodes** into the organ.

Example: Patient presents with abdominal adenopathy. Surgical exploration documents direct invasion of the stomach from the regional lymph nodes. Code abdominal lymph nodes (C772).

Rule PH35 Code the primary site to the organ when lymphoma is present in an **organ** and that organ's **regional lymph nodes.**

Note: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes.

Example 1: Lymphoma is present in the kidney and peri-renal lymph nodes. Code the primary site to kidney (C649).

Example 2: Lymphoma is present in the stomach and the gastric lymph nodes. Code the primary site to stomach, NOS (C169).

Example 3: Lymphoma is present in the spleen and the splenic lymph nodes. Code the primary site spleen (C422).

Rule PH36 Code the primary site to lymph nodes, NOS (C779) when **lymphoma** is present in an **organ(s)** and **lymph nodes** that are not **regional** for that organ and the **origin cannot be determined** even after consulting the physician.

Note 1: Lymphoma can spread from organs to **regional** lymph nodes, but does **not** spread from the organ directly to **distant** lymph nodes

Example: The patient has positive mediastinal (C771) and cervical (C770) lymph nodes and involvement of the stomach (C169). No further information is available. Code to lymph node, NOS (C779).

Note 2: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes.

Note 3: See Appendix C for help in identifying lymph node names, chains, regions, and codes.

Rule PH37 Code primary site to unknown primary site (C809) only when there is no evidence of lymphoma in lymph nodes AND the physician documents in the medical record that he/she suspects that the lymphoma originates in an organ(s). See ICD-O-3 Rule D.

When this module does not apply to the case being abstracted, go to Module 8.

Module 8: Histology Rules Only: All hematopoietic and lymphoid neoplasms 9590/3-9992/3
PH38 - PH39

Rule PH38 Code the non-specific (**NOS**) histology when the diagnosis is

- **One non-specific histology AND**
- **Two or more specific histologies AND**
- The hematopoietic DB multiple primaries calculator documents the specific histologies and NOS are the **same primary AND**
- No further information is available

Note 1: Use Appendix E: Histology "NOS" Tables to identify the NOS histologies.

Note 2: Use the Hematopoietic DB multiple primaries calculator to confirm that the NOS and specific histologies are the same primary.

Example: The diagnosis is myeloproliferative disorder, NOS (9960/3), polycythemia vera (9950/3), essential thrombocythemia (9962/3). The Hematopoietic DB multiple primaries calculator shows myeloproliferative disorder and polycythemia vera are the same primary. The multiple primaries calculator also shows myeloproliferative disorder and essential thrombocythemia are the same primary. Follow-back produces no additional information. Code the histology myeloproliferative disorder, NOS (9960/3).

Rule PH39 Code the specific histology when the diagnosis is

- One non-specific (NOS) histology AND
- One specific histology AND
- The Hematopoietic DB multiple primaries calculator documents the specific histology and NOS are the same primary

Note 1: Use Appendix E: Histology "NOS" Tables to identify the NOS histologies.

Note 2: Use the Hematopoietic DB multiple primaries calculator to confirm that the NOS and specific histology are the same primary.

When this module does not apply to the case being abstracted, go to Module 9.

Module 9: Default Rules: All hematopoietic and lymphoid neoplasms 9590/3-9992/3
PH40-PH41

Rule PH40 Use the **Hematopoietic DB** to determine the primary site and histology when rules PH1-PH39 do **not** apply.

Rule PH41 When the histology code cannot be determined using the Hematopoietic DB, code the histology with **the numerically higher** ICD-O-3 code.

This is the end of the rules for coding primary site and histology.

Grade of Tumor Rules Text Format

Note 1: Use the Grade of Tumor Rules (G1-G11) to assign the correct code in the grade field.
Note 2: Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade.

Rule G1 Code cell type not determined, not stated, not applicable (code 9) for the following myeloproliferative neoplasms, myeloproliferative/myelodysplastic syndromes, myelodysplastic syndrome, histiocytic and dendritic cell neoplasms

9740/3	9946/3
9741/3	9950/3
9742/3	9961/3
9751/3	9962/3
9755/3	9963/3
9757/3	9964/3
9758/3	9975/3
9759/3	9980/3
9801/3	9982/3
9805/3	9982/3
9806/3	9983/3
9807/3	9985/3
9808/3	9986/3
9809/3	9989/3
9875/3	9991/3
9876/3	9992/3
9945/3	

Note 1: These neoplasms do not have a specific codable phenotype
Note 2: See Tables B1, B3, B4, and B11 in Appendix B for neoplasm terms and codes.

Rule G2 Use statements from **any** part of the medical record including, but not limited to

- Pathology report **OR**
- History and physical **OR**
- Consultation **OR**
- Final diagnosis **OR**
- Face sheet

Rule G3 Use codes 5, 6, 7, 8, and/or 9 **only** -- even if there is a statement giving the cell type in the medical record

Note 1: Do **not** code descriptions "low grade," intermediate grade," or "high grade" in the Tumor Grade field. These terms refer to the Working Formulation categories of lymphoma diagnosis.

Note 2: Do **not** code the descriptions "Grade 1," "Grade 2," or "Grade 3" in the Tumor Grade field. These grades represent histology types of lymphoma rather than differentiation.

Rule G4 Code T-cell (**code 5**) for the following neoplasms; **T-cell** is part of the neoplasm name or the neoplasm is of **T-cell origin**

9701/3
9702/3
9705/3
9708/3
9709/3
9714/3 (unless pathologist specifically designates as a B-cell)
9716/3
9717/3
9718/3
9724/3
9725/3
9726/3
9827/3
9831/3
9834/3
9837/3

Note 1: Record T-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention T-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.

Note 2: When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Rule G5 Code T-cell (code 5) when the neoplasm is identified as **T-cell, T-cell phenotype, T-precursor, Pre-T, gamma-delta-T, or null-cell T-cell**

Rule G6 Code B-cell (code 6) for the following **B-cell precursor lymphoid neoplasms** and the **mature B-cell neoplasms**

9591/3	9684/3
9596/3	9687/3
9597/3	9688/3
9670/3	9689/3
9671/3	9690/3
9673/3	9691/3
9678/3	9695/3
9679/3	9698/3
9680/3	9699/3(continued on next page)

9712/3	9813/3
9728/3	9814/3
9731/3	9815/3
9732/3	9816/3
9734/3	9817/3
9737/3	9818/3
9738/3	9823/3
9762/3	9833/3
9811/3	9836/3
9812/3	9940/3

Note 1: Record B-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention B-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.

Note 2: When the medical record or pathology report contains one of these terms with a different phenotype (T-cell, null-cell, or NK-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Note 3: See Tables B7 and B8 in Appendix B.

Rule G7 Code B-cell (**code 6**) when the disease is identified as **B-cell, B-cell phenotype, B-precursor, pre-B, or null-cell and B-cell.**

Rule G8 Code Null cell, non-T non-B (code 7) when the disease is described as **null cell, non-T non-B,** or **common cell.**

Rule G9 Code **NK-cell** (natural killer cell) (code 8) for the following neoplasms; NK-cell is a part of the **neoplasm's name**

9719/3
9948/3

Note 1: Record NK-cell even though it is not mentioned as a specific phenotype in the pathology or other test report(s). Frequently physicians do not mention NK-cell phenotype because they know the phenotype or they understand that the phenotype is inherent in the disease classification/name.

Note 2: When the medical record or pathology report contains one of these terms with a different phenotype (B-cell, T-cell, or null-cell) check with the pathologist to determine whether the disease name is correctly recorded. It is possible that the mention of a different phenotype may be the result of the pathologist using a different disease classification.

Note 3: See Table B9 in Appendix B.

Rule G10 Code Natural Killer (NK) cell (**code 8**) when the disease is described as **NK cell, natural killer cell, nasal NK/T-cell lymphoma, or null-cell and NK cell.**

Rule G11 Code cell type not determined, not stated, not applicable (code 9) when

- There is **no statement describing the cell type OR**
- The cell type is described as **combined T AND B cell OR**
- The cell type is described as **combined B AND NK cell**

Note: There is a new site-specific factor to collect combination cell types for hematopoietic or lymphatic neoplasms in the Collaborative Stage Data Collection System, Version 2.

Glossary

Allogeneic bone marrow/stem cell transplant: Marrow is used from a donor whose Human Leukocyte Antigens (HLA) closely match the patient's. Stem cells are taken either by bone marrow harvest or by apheresis from a genetically matched donor. (See also Bone marrow transplant and Stem cell transplant.).

Anemia: The number of red blood cells is below normal for blood count.

Apheresis: Process in which the blood of a donor or patient is passed through an apparatus that separates out one particular constituent and returns the remainder to circulation.

Autologous bone marrow/stem cell transplant: The patient's own bone marrow is used. (See also Bone marrow transplant and Stem cell transplant.)

B-cells: B-cells make antibodies against antigens and are produced in the bone marrow and mature in the bone marrow. B cells are lymphocytes that play a large role in the humoral immune response (as opposed to the cell-mediated immune response, which is governed by T-cells).

B-cell and T-cell (combined): Because a major loss or dysfunction of T cells can cause secondary B-cell deficiency, a number of disorders show clinical manifestations of combined B- and T-cell deficiency, though the only pathology is in the T-cell. In contrast, some diseases appear to primarily involve the T-cells and do not appear to affect antibody production. Those diseases are discussed in T-cell disorders.

B-cell leukemia: A type of cancer that forms in B-cells. This neoplasm usually occurs in adults and may be indolent (slow growing) or aggressive (fast growing). Many different types of B-cell lymphomas and leukemias have been identified. (See B-cells.)

B-cell lymphoma: A type of cancer that forms in B-cells. B-cell lymphomas usually occur in adults and may be either indolent (slow-growing) or aggressive (fast-growing). There are many different types of B-cell lymphomas, and prognosis and treatment depend on the type and stage of cancer. (See B-cells.)

BCR-ABL cancer gene: A mutant gene that is formed when a piece of chromosome 9 attaches to the end of chromosome 22. The BCR-ABL cancer gene gives the cell instructions to make a protein that leads to CML.

Bence Jones protein: A protein made by myeloma cells that is found in the plasma and urine of many patients with myeloma. This type of protein is also called "light chains" because it represents a smaller segment of the whole immunoglobulin molecule, composed of heavy and light chains.

Biopsy: The removal of a sample of tissue for purposes of diagnosis. (Many definitions of "biopsy" stipulate that the sample of tissue is removed for examination under a microscope. This may or may not be the case. The diagnosis may be achieved by other means such as by analysis of chromosomes or genes.)

Blast cell: Immature blood-forming cell.

Blood chemistry tests: These measure the amount of certain chemicals in the blood. They are not used to diagnose leukemia; rather, they detect liver or kidney damage caused by leukemic cells or the chemotherapy drugs.

Blood count (CBC or complete blood count): A count of the number of red blood cells, white blood cells and/or platelets in a given sample of blood. (See also peripheral blood smear).

Bone marrow aspiration: The removal of a small amount of bone marrow (usually from the hip) through a needle. The needle is placed through the top layer of bone and a liquid sample containing bone marrow cells is drawn into a syringe. The sample is then sent for cytologic examination. Bone marrow aspiration is done to diagnose and follow the progress of various conditions, including anemia and cancer, and to obtain marrow for transplantation.

Bone marrow biopsy: The removal of a sample of bone marrow and a small amount of bone (usually from the hip) through a large needle. The sample is a core biopsy to obtain bone marrow together with bone fibers. The sample is examined under a microscope (pathology) to see the cells and architecture of the bone marrow.

Bone marrow transplant: A complex treatment that may be used when cancer is advanced or has recurred. The bone marrow transplant makes it possible to use very high doses of chemotherapy that would otherwise be impossible because they destroy the patient's normal marrow cells as well. When used for advanced or recurrent cancer, a portion of the patient's or a donor's bone marrow is withdrawn, purged of tumor cells if possible (in the case of autologous transplantation), and stored. Then the patient is given high doses of chemotherapy to kill the cancer cells. But the drugs also destroy the remaining bone marrow, thus robbing the body of its natural ability to fight infection. The stored marrow is given by transfusion (transplanted) to rescue the patient's immune defenses. Currently most patients are treated with stem cell transplants rather than bone marrow transplants.

Bone marrow: The soft tissue in the internal portion of flat bones and vertebrae that produces new blood cells.

CBC: (See blood count.)

CD; **cluster of differentiation**: Used to define the findings in immunophenotyping (See definition of immunophenotyping). The CD is the antigen found in the cell and is associated with a number such as CD16 which may be present on leukemic lymphoblasts.

Chromosome: A part of the cell that carries genes. Genes give instructions that tell the cell what to do. The cell has 46 chromosomes, 22 pairs of chromosomes plus two sex chromosomes. Females have two "X" chromosomes. Males have one "X" and one "Y" chromosome.

Chromosome abnormality or mutation: Chromosomal anomalies may reflect an atypical number of chromosomes (karyotype) or a structural abnormality in one or more chromosomes. Chromosome anomalies usually occur when there is an error in cell division following mitosis or meiosis. Many types of chromosome anomalies exist, including single chromosome mutations such as deletion, duplication and inversion. The two major chromosome mutations include: insertion or translocation of genes.

Chromosome deletion: A portion of the chromosome is missing or deleted.

Chromosome duplication: A portion of the chromosome is duplicated, resulting in extra genetic material.

Chromosome insertion: Insertions can vary in size from one base pair incorrectly inserted into a DNA sequence to a large section of one chromosome inserted into another chromosome. Basically, it is the loss of a portion of one chromosome with the "lost" portion inserted into a different chromosome.

Chromosome inversion: A portion of the chromosome has broken off, turned upside down, and reattached; therefore the genetic material is inverted. In contrast to chromosome insertion where a different chromosome is inserted into another chromosome, chromosome inversion uses the same chromosome. Basically it is a switch of a small portion of one chromosome with a small portion of another chromosome so all of the genetic material is retained, but the two small portions of chromosomes have changed places with each other.

Chromosome translocation: Translocation occurs when a portion of one chromosome is transferred to another chromosome. There are two main types of translocations; reciprocal and Robertsonian. In a reciprocal translocation, segments from two different chromosomes have been exchanged. In a Robertsonian translocation, an entire chromosome has attached to another chromosome at the centromere.

Chromosome: A part of the cell that carries genes. Genes give instructions that tell the cell what to do. The cell has 46 chromosomes, 22 pairs of chromosomes plus two sex chromosomes. Females have two "X" chromosomes. Males have one "X" and one "Y" chromosome.

Clinical diagnosis: A diagnosis made on the basis of physician expertise; there is no microscopic or imaging confirmation. For hematopoietic and lymphatic neoplasms, clinical diagnosis includes diagnoses of exclusion where microscopic and imaging confirmation is equivocal and the physician uses his/her expertise to evaluate the clinical presentation and equivocal tests to make a diagnosis.

Colony-stimulating factors (CSF): Types of growth factors that promote growth and division of blood-producing cells in the bone marrow. CSFs are naturally produced in the body. Note: CSF agents are classified as immunosuppressing drugs, but are not coded on the cancer abstract.

Combined modality therapy: Two or more types of treatment used alternately or together to improve results. For example, surgery for cancer is often followed by chemotherapy to destroy any cancer cells that may have spread from the original site, or local radiation is given to treat any cancer cells that may have been left behind after surgery.

Complete remission: A term that is applied to a patient's health status after treatment, when there is no sign of the disease using standard tests specific for that disease and the patient has returned to good health.

Consolidation therapy: A term usually applied to the treatment of acute leukemia for drug treatment given to patients in remission after induction therapy. The aim of consolidation therapy is to kill as many of the remaining cancer cells as possible.

Cutaneous lymphoma: Lymphoma originating in the skin.

Cytochemistry: After cells are placed on slides they are exposed to chemical stains (dyes) that are attracted to or react with some types of leukemia cells.

Cytogenetic analysis: The term for a lab test that is used to examine the chromosomes in marrow, blood, and lymph node cells. It can confirm that the cells are cancer cells (malignant) and in some cases the results may guide the intensity of therapy.

Cytogenetic response (cytogenetic remission): A treatment response in which there are no leukemia, lymphoma or myeloma cells detected in the blood and/or marrow by the FISH test.

Cytogenetics: The study of chromosomes, the visible carriers of DNA. This is a fusion science joining cytology, the study of cells, with genetics, the study of inherited variation.

Cytokines: Products of cells of the immune system that regulate the immunologic, inflammatory and reparative responses. Some may stimulate immunity and cause the regression of cancers.

Cytostatic: Describes the way some anti-cancer drugs work, not what type of drug they are. Cytostatic treatments stop the cancer cells from multiplying; they do not kill cancer cells.

Cytotoxic: Toxic to cells; cell-killing. Cytotoxic describes any agent or process that kills cells. Chemotherapy and radiotherapy are forms of cytotoxic therapy.

Definitive diagnosis: For the purpose of these rules, the definitive diagnosis is the diagnostic method or methods listed in the Hematopoietic DB (under the category "definitive diagnostic method"). The definitive diagnoses are the methodologies used to identify the specific neoplasm.

Dendritic cell: The dendrites are immune cell with threadlike tentacles which enmesh antigen; T cells then attack the antigen. Both Langerhans cells, found in the skin and follicular dendritic cells, found in lymphoid tissues, and are types of dendritic cells.

Differentiation: The normal process through which cells mature so they can carry out the jobs they were meant to do. Cancer cells are generally less differentiated than their normal counterparts. Differentiation also refers to the greater or lesser degree of morphologic similarity that cancer cells have to normal cells.

DNA: Abbreviation for deoxyribonucleic acid. DNA holds genetic information on cell growth, division, and function.

Donor lymphocyte infusion: A treatment that uses an infusion of white cells called lymphocytes from the original stem cell donor.

Drug resistance: When a drug used to treat a patient's disease does not work or stops working.

Electrophoresis, serum, protein, fluorescent and urine: A type of test that separates substances, especially proteins, and analyzes molecular structure. Electrophoresis may be used to provide a confirmation of a diagnosis of multiple myeloma. Electrophoresis is used to detect M proteins in the urine or serum (M-spike).

Epstein Barr Virus (EBV): Discovered by Epstein, Achong, and Barr by electron microscopy from Burkitt lymphoma tissue. EBV is also called Human herpes virus 4 (HHV-4). EBV is a precursor to several kinds of cancer.

Etiology: The cause of a disease. Cancer has many causes, although research is showing that both genetics and lifestyle are major factors in many cancers.

Extramedullary myeloma: A tumor of plasma cells in a site other than bone marrow.

Extramedullary: Occurring outside any of the medullas including the medulla oblongata and the medullary cavities of the bones.

Extraosseous: Occurring outside of the bone or bones.

FISH: Analysis can be done on blood, bone marrow, lymph nodes, and any other tissue. The short name for a test called "fluorescence in situ hybridization," a test to measure the presence in cells of a specific chromosome or gene. This test can be used to plan treatment and to measure the results of treatment.

Flow cytometry: A method of counting types of cells with fluorescent tags on the surfaces of the cells. Flow cytometry is often used to determine the type of leukemia, lymphoma or myeloma cells that are present. For example, each disease subtype has a specific pattern of markers on its cell surface. Flow cytometry can also detect residual levels of disease after treatment.

Gene: Parts of cells that give instructions for making proteins. Proteins help the cell do its job.

Genetic testing: Analysis can be done on blood, bone marrow, lymph nodes, and any other tissue. Laboratory studies of blood or other tissue to analyze DNA in order to identify chromosome abnormalities which identify specific neoplasms. Genetic testing is also done to Identify genetic alternation that may indicate an increased risk for developing a specific disease or disorder.

Genotype: The genetic makeup, as distinguished from the physical appearance, of an organism or a group of organisms.

Hematologic response (hematologic remission): A treatment response where the leukemia, lymphoma or myeloma cell numbers are decreased in the blood; and, red cell count, white cell count, and platelet count are either at or near normal values.

Hematologist: A physician who treats blood cell diseases.

Hematopoietic: Pertaining to hematopoiesis which means the production of all types of blood cells.

Hematopoietic Neoplasms (as used in these rules): Includes leukemias, lymphomas, and mast cell tumors (9590-9992).

Hodgkin lymphoma (HD), Hodgkin disease (HD), Hodgkins: A cancer of the lymphatic system. Hodgkin lymphoma appears to originate in a lymph node and later spreads to the spleen, liver, and bone marrow. The presence of the Reed-Sternberg cell distinguishes Hodgkin lymphoma from non-Hodgkin lymphoma.

Human leukocyte antigens (HLA): HLA tissue typing identifies the best tissue/blood cell match between donors and recipients.

ICD-O: The International Classification of Diseases for Oncology published by the World Health Organization (WHO). This reference is used to code the primary site and histology for reportable neoplasms in a standardized manner. These codes allow historic and current data to be grouped for reporting and analysis.

Immune response: The reaction of the body to foreign material such as an infection-causing microorganism, an immunization, or the cells of another individual used for an allogeneic stem cell transplant.

Immune system: The complex system by which the body resists infection by microbes such as bacteria or viruses. The immune system may also help the body fight some cancers. It is also responsible for the rejection of transplanted tissues or organs.

Immunocytochemistry: Cells from the bone marrow are treated with special antibodies that cause certain types of cells to change color. Immunocytochemistry is sometimes helpful in determining the exact type of acute leukemia. It is not necessary in most cases of chronic leukemia.

Immunophenotyping: Analysis can be done on blood, bone marrow, lymph nodes, and any other tissue. A sample of blood, bone marrow cells, or lymph node cells is analyzed to determine the types of antigens or markers on the surface of the cell. This analysis is used to diagnose specific types of leukemia and lymphoma, for example, myelogenous leukemic cells can be distinguished from lymphomatous leukemic cells. The antigen in the cell is usually referred to as CD followed by a number (See definition of CD).

Immunosuppression: A state in which the body's immune system does not respond as it should. This condition may be present at birth, or it may be caused by infections (such as human immunodeficiency virus (HIV)) or by cancer therapies such as cancer-cell killing (cytotoxic) drugs, radiation, and bone marrow transplantation.

Induction therapy: The initial treatment of a patient with a blood cancer with chemotherapy (or radiation therapy). The aim of induction therapy is to kill a maximum number of blood cancer cells so as to induce a remission (absence of signs or effects of the disease).

JAK2, Janus kinase 2, Exon 12: JAK2 is a gene mutation that increases susceptibility to several myeloproliferative neoplasms (MPNs). Testing for the JAK2 mutation is done on whole blood or on bone marrow. Nearly all people with polycythemia vera and about half of those with essential thrombocythemia and primary myelofibrosis have the mutation. When JAK2 is positive, the MPN is definitely reportable; however JAK2 does not identify which specific MPN is present. Correlation for other clinical findings and laboratory findings is needed to determine the specific histology. The JAK2 mutation has also been found in a minority of patients with other myeloid stem cell disorders, including chronic myelomonocytic leukemia.

Karyotyping: To arrange and classify chromosomes based on number, size, shape, and other characteristics.

Late effects: Medical problems that do not develop or become apparent until months or years after treatment ends. Examples of late effects include the development of a treatment-related cancer or heart disease.

Leukapheresis: A process in which extra white cells are removed by a machine.

Leukemia: Cancer of the blood or blood-forming organs. People with leukemia often have a noticeable increase in white blood cells (leukocytes) in the peripheral blood.

Leukocyte alkaline phosphatase (LAP) test: Useful in distinguishing CML from other types of leukemia and benign conditions.

Leukopenia: Decrease in the while blood cell count, often a side effect of chemotherapy.

Light chains: A part of the monoclonal (M) protein in myeloma. Monoclonal immunoglobulin (protein), like normal immunoglobulin is usually made up of two heavy (larger) chains and two light (smaller) chains attached to each other. The abnormal production of immunoglobulin protein by myeloma cells sometimes results in parts of the molecule that is the heavy chain or light chain, being made and discharged from the myeloma cells. They can each be measured in plasma. The light chains are small enough to pass through the kidney and enter the urine, where they can be detected.

Long-term effects: Medical problems that persist for months or years after treatment ends, for example, infertility, growth problems in children, or cancer treatment-related fatigue.

Lymph nodes (lymph glands): Small bean-shaped collections of immune system tissue such as lymphocytes and macrophages found along lymphatic vessels. The nodes remove cells and cell waste from lymph and help fight infections.

Lymph: Clear fluid that flows through the lymphatic vessels and contains cells known as lymphocytes. These cells are important in fighting infections and may also have a role in fighting cancer.

Lymphatic organ: An organ of the immune system where lymphocytes develop and congregate such as tonsils, adenoids, thymus and spleen. (There is disagreement among the experts on the division of lymphatic organs and lymphatic tissues. For the purpose of these rules, we use them as synonyms meaning any aggregate of lymph cells within the body).

Lymphatic system: The tissues and organs, including the bone marrow, spleen, thymus, and lymph nodes, that produce and store cells that fight infection and disease.

Lymphatic tissue: Immune system tissue where lymphocytes develop and congregate such as tonsils, adenoids, thymus and spleen. (There is disagreement among the experts on the division of lymphatic organs and lymphatic tissues. For the purpose of these rules, we use them as synonyms meaning any aggregate of lymph cells within the body).

Lymphatic vessels: These channels connect the lymph nodes. They contain lymph - a fluid that carries lymphocytes as they circulate from one lymph node area to another. The lymphatic channels are connected to the blood vascular system permitting lymphocytes to enter the blood.

Lymphoblastic: A term used to describe a type of blood cell disease caused by young or immature lymphocytes or "lymphoblasts". An example is acute lymphoblastic leukemia, which is characterized by the presence of malignant (cancerous) lymphoblasts (immature lymphocytes).

Lymphocyte: A type of white blood cell that is part of the immune system. There are three major types of lymphocytes: B lymphocytes that produce antibodies to help combat bacterial, fungal or viral infections; T lymphocytes that have several functions, including assisting B lymphocytes to make antibodies; and natural killer (NK) cells that can attack virus-infected cells or tumor cells.

Lymphocytic: A term used to describe a type of blood cell abnormality caused by lymphocytes. An example is chronic lymphocytic leukemia, which is characterized by the presence of malignant (cancerous) lymphocytes. Sometimes used as a synonym for "lymphoblastic."

Lymphoma: A type of cancer that begins with a malignant change in a lymphocyte, lymph node cell or a cell in the lymphatic tissue of the marrow, gastrointestinal tract, spleen, skin or other sites.

M protein (monoclonal protein): Myeloma cells make a protein called monoclonal immunoglobulin, sometimes referred to as M protein. M protein, like normal immunoglobulin is usually made up of two heavy (larger) chains and two light (smaller) chains attached to each other. The amount of M protein in the blood can be measured in the laboratory. It is used to estimate the extent of the myeloma and to follow the effects of treatment.

Marrow: The spongy center inside of bones.

Matched donor: A person whose major tissue types are identical to those of a patient who is seeking a stem cell transplant. The patient can be given the donor's healthy matched stem cells, which can restore blood and immune cells after high-intensity cancer treatment.

Molecular genetic studies: The branch of genetics that deals with hereditary transmission and variation on the molecular level.

Molecular response: A treatment response is called a complete molecular remission if no leukemia cells in the blood and/or marrow can be detected by PCR.

Monoclonal antibody therapy: A type of therapy that targets and kills cancer cells. Monoclonal antibodies are immune proteins made in the laboratory. They are designed to target to a specific blood cancer cell. They produce less toxic effects on normal tissues than chemotherapy.

Monocyte: A type of white cell. Monocytes and neutrophils are the two major microbe-eating and killing cells in the blood.

Multiple myeloma (MM), myeloma, Kahler disease, myelomatosis, plasma cell myeloma: A type of cancer that begins in plasma cells (white blood cells that produce antibodies). MM is characterized by excessive numbers of abnormal plasma cells in the bone marrow and overproduction of intact monoclonal immunoglobin.

Myelofibrosis: A progressive disease of the bone marrow. Neoplastic bone marrow stem cells lodge and grow in multiple sites outside the bone marrow. Symptoms include enlargement of the spleen and a gradual replacement of the bone marrow elements by fibrosis (scarring), progressive anemia, and variable changes in the number of white blood cells and platelets.

Myelogenous: A term used to describe a form of blood cancer that begins in a marrow stem cell or early marrow progenitor cell. A blood cancer that begins in the marrow is called leukemia. Myelogenous leukemias usually do not directly affect lymphocytes. The terms "myeloid" or "myelocytic" are sometimes used instead of "myelogenous."

Myeloma: A tumor of antibody-producing cells, called plasma cells, which are normally found in the bone marrow.

Natural killer cell (NK cell): NK cells, like killer T cells, attack and kill cancer cells and cells infected by microorganisms. NK cells can react against and destroy another cell without prior sensitization to that cell. Natural killer (NK) cells are part of our first line of defense against cancer cells and virus-infected cells. NK cells are small lymphocytes that originate in the bone marrow.

Neutropenia: A decrease below normal in the concentration of neutrophils, a type of white cell.

Neutrophil: A type of white cell. Neutrophils and monocytes are the two main microbe-eating cell and infection-fighting in the blood.

Non-Hodgkin lymphoma (non-Hodgkins, NHL): A cancer of the lymphatic system. What distinguishes non-Hodgkin lymphoma from Hodgkin lymphoma (Hodgkin disease) is the absence of a type of cell called the Reed-Sternberg cell. This cell is present only in Hodgkin lymphoma.

Nonmyeloablative stem cell transplant (mini-transplant): A type of allogeneic stem cell transplant that does not use high-dose chemotherapy as a treatment. The patient takes special drugs so that his or her immune system does not reject the transplanted stem cells. Over a long time, the donated cells replace the patient's blood and immune system cells. The donated cells also attack the leukemia, lymphoma or myeloma cells. Other drugs to help the transplanted stem cells fight the blood cancer without attacking healthy cells are being tested in clinical trials.

Nucleus: A part of the cell containing the chromosomes and genes.

Null cell: A lymphocyte that develops in the bone marrow and lacks the characteristic surface markers of the B and T lymphocytes. A null cell is a large granular lymphocyte without surface markers or membrane-associated proteins from B lymphocytes or T lymphocytes. Natural killer cells are usually null cells with surface marker CD 16, which binds to the Fc portion of the IgG, thereby destroying it.

Oncologist: A physician who treats patients with cancer.

Patch mycosis fungoides: The early stage of mycosis fungoides is called the “patch stage.” The skin is infiltrated by patches or lumps of lymphocytes (white cells). In this early stage, the neoplasm appears in ovoid patches that do not cover large amounts of skin. During the patch stage of mycosis fungoides it is difficult to distinguish the neoplasm from eczema or psoriasis.

Pathologist: A doctor who examines cells and tissues obtained from biopsies to determine the type of disease present.

PCR: The short name for a lab test called "polymerase chain reaction," a very sensitive test that can measure the presence of a blood cancer cell marker in the blood. It is used to detect remaining blood cancer cells that are below the detection of cytogenetic methods (e.g. FISH).

Peripheral blood smear: Blood is viewed under a microscope to count different circulating blood components (red cells, white cells, platelets, etc.) to see whether the cell population level is normal. (See also Blood count.)

Phenotype: The observable traits or characteristics of an organism, for example hair color, weight, or the presence or absence of a disease. Phenotypic traits are not necessarily genetic.

Plasma: Part of the blood that is mostly water, with some vitamins, minerals, proteins, hormones and other natural chemicals.

Plasmacytoma, extraosseous/extramedullary: Localized plasma cell neoplasms that arise in tissue other than bone.

Plasmacytoma, solitary plasmacytoma of bone/osseous plasmacytoma: Localized bone tumor consisting of monoclonal plasma cells.

Platelet (thrombocyte): A type of blood cell that prevents bleeding and forms a plug that stops bleeding after an injury to the body.

Ploidy: Degree of repetition of the base number of chromosomes in a cell. A diploid cell has double the base number, one set from each parent, while a monoploid cell has only one set (a haploid is a sex cell, which also has only one set). Aneuploidy is an abnormal condition in which a chromosome is missing or an extra one is inserted.

Polymerase chain reaction (PCR): PCR is a molecular technique which allows the production f large quantities of a specific DNA from a DNA template.PCR has transformed the way that most studies requiring the manipulation of DNA fragments and DNA cloning may be performed.

Port: An implanted port is a type of long-term catheter. The port is surgically inserted under the skin's surface on the upper chest wall. After the site heals, no dressings are needed and no special home care is required. When medicines are needed, a physician, physician assistant or nurse inserts a needle through the skin to access the port. The patient can choose to have a local numbing cream applied to the injection site before the port is used. Blood can be drawn, and blood products can be received through this device.

Primary cutaneous B-cell lymphoma (PCBCL): B-cell lymphoma originating in the skin. There is no evidence of extracutaneous involvement at the time of diagnosis or for six months after diagnosis. Two subtypes of PCBCL have been identified by the European Organization for Research and Treatment of Cancer: follicular center cell lymphoma and large B-cell lymphoma.

Protocol: A formal outline or plan, such as a description of what treatments a patient will receive and exactly when each should be given and in what doses.

Radiation therapy: The use of x-rays or other high-energy rays to kill cancer cells.

Radioimmunotherapy: A treatment that uses antibodies to carry a radioactive substance to cancer cells to kill them. They are used in the treatment of lymphoma and lymphocytic leukemia.

Red cell: A blood cell that carries oxygen and delivers it to the body.

Reed Sternberg cells: Giant cells found on light microscopy in biopsies from patients with Hodgkin lymphoma and certain other disorders. They are derived from B lymphocytes. They are named after Dorothy Reed Mendenhall and Carl Sternberg who provided the first definitive microscopic descriptions of Hodgkin lymphoma.

Refractory disease: Disease that does not respond to therapy.

Relapse or recurrence: When disease comes back after it has been successfully treated.

Remission: A period of time with no signs of disease and/or when the patient does not have any symptoms of the disease.

Richter's transformation, Richter's syndrome (RS): Transformation of B-cell chronic lymphocytic leukemia (CLL) or hairy cell leukemia (HCL) to diffuse large B-cell lymphoma.

Risk factor: A factor that may increase the chance that a person will develop a disease or condition. For example, cigarette smoking is a risk factor for lung cancer.

Side effect: The signs or symptoms a patient may have from the effects of treatment on healthy cells.

Signs and symptoms: A sign is a change in the body that the doctor sees in an exam or a lab test. A symptom is a change in the body that a patient can see or feel.

Skin infiltration: For lymphoma diagnoses, skin infiltration means involvement of the skin (not spread to the skin). The term "skin infiltration" is commonly used with cutaneous lymphomas. Cutaneous lymphomas originate in the skin.

Smoldering leukemia, preleukemia, myelodysplastic syndrome: The myelodysplastic syndromes are a group of diseases in which the bone marrow does not make enough healthy blood cells.

S-phase fraction (SPF): The percentage of cells that are replicating their DNA. DNA replication usually means that a cell is getting ready to divide into two new cells. In a tumor, a low SPF is a sign that a tumor is slow-growing; a high SPF shows that the cells are dividing rapidly and the tumor is growing quickly.

Spleen: The spleen is a lymphatic organ which is encased in a thick capsule. Within the spleen are two types of tissue: the white pulp, which is a lymphoid tissue and usually surrounds the blood vessels, and the red pulp, which is a network of channels (sinuses) that are filled with blood. The white pulp has typical lymphoid elements such as plasma cells, lymphocytes, and lymphatic nodules, which help fight infection. The red pulp destroys old red blood cells and filters the blood.

Stem cell: A type of cell found in marrow that makes red cells, white cells and platelets.

Stem cell transplant: A variation of bone marrow transplantation in which immature blood cells called stem cells are taken from the blood of the patient or a donor. Later, in the lab, the cells are stimulated with growth factors to produce more stem cells that are then transfused to the patient. Most hematopoietic diseases are treated with stem cell transplants rather than a bone marrow transplant.

Syngeneic bone marrow transplant: Transplant in which an identical twin is the bone marrow donor.

Systemic therapy: Treatment that reaches and affects cells throughout the body; for example, chemotherapy.

T cells: One type of white blood cell that attacks virus-infected cells, foreign cells, and cancer cells. T cells also produce a number of substances that regulate the immune response. T cells are produced in the bone marrow and mature in the thymus. They are the most common type of lymphocyte, itself divided into at least three subpopulations on the basis of function: cytotoxic, or killer, T lymphocytes; helper T lymphocytes; and suppressor T-lymphocytes.

T-cell lymphoma: A disease in which lymphoid cells called T-cells (or T lymphocytes) becomes malignant. T-cell lymphomas account for a minority (about 15%) of non-Hodgkin lymphomas in the US. The T-cell lymphomas are highly diverse and include lymphoblastic lymphoma (mainly in children and adolescents), peripheral T-cell lymphoma (a heterogeneous group of generally aggressive diseases), mycosis fungoidies (called Sezary syndrome if the malignant T cells circulate in blood), and anaplastic large cell lymphoma (ALCL), which can be primary cutaneous ALCL or systemic ALCL. T-cell lymphomas account for about half of pediatric lymphomas.

Thrombocyte: Another word for platelet.

Thrombocytopenia: A decrease below normal in the number of platelets.

Vaccine therapy: A type of treatment under study for leukemia, lymphoma, or myeloma. This type of vaccine would not prevent the disease. The vaccine would increase the immune system's attack against cancer cells that remain after treatment with drugs.

Waldenstrom macroglobulinemia: A clinical diagnosis, not a pathology diagnosis. Most cases are associated with lymphoplasmacytic lymphoma, but occasionally it is associated with other subtypes of NHL. This is the only hematopoietic disease coded to blood (C420).

Watch and wait: An approach in which a physician closely observes a patient's condition with periodic medical exams and lab tests, without giving drugs or other forms of treatment for the disease in question. For some patients with non-growing or very slow-growing disease and no symptoms, watch and wait may be preferred; it allows the patient to avoid drug treatment and its potential side effects until drugs are needed. This approach is based on studies that indicate early treatment in the specific situation in question is not beneficial.

White cell: Type of cell that fights infections in the body. There are two major types of white cells: germ-ingesting cells (neutrophils and monocytes) and lymphocytes, which provide an immune response to infection.

World Health Organization (WHO): The directing and coordinating authority for health within the United Nations. It is responsible for providing leadership on global health matters, shaping the health research agenda, setting norms and standards, articulating evidence-based policy options, providing technical support to countries, and monitoring and assessing health trends (from http://www.who.int/about/en/).

Appendix A
History of Hematopoietic and Lymphoid Neoplasm Coding

History of Coding Diseases of Lymphoid tissue and Hematopoietic systems

Historically, diseases of lymphoid tissues and the hematopoietic system were believed to be separate entities, and the coding structure of the International Classification of Diseases was developed with this in mind. Prior to the early 1990s, the classification systems for lymphomas described malignant cells by their morphologic characteristics; for example, the size and shape of the tumor cell and its pattern of tumor growth and spread. The *International Classification of Diseases for Oncology,* Third Edition (ICD-O-3) says this about the historic classifications:

> Over the past 50 years many classifications of leukemia and lymphoma have been proposed. Some of these had a major impact on clinical practice while others are now largely forgotten. For most of this period, however, the distinction between lymphoma and leukemia has been regarded as of fundamental importance and classifications have tended to evolve separately (ICD-O-3, p. 13).

The World Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues, 4th Edition, was published in 2008. The 4th Edition of this world-renowned reference describes the current standard classification system for tumors of the hematopoietic and lymphoid systems. The 2008 classification continues to be based on the principles originally outlined in the REAL classification system (grouping by phenotype). These principles have now been applied to the classification of myeloid, lymphoid, mast cell, and histiocytic/dendritic neoplasms. Additionally, when specialized testing demonstrates one or more disease-specific or disease-defining characteristic using immunophenotyping and/or genetic testing, these characteristics have been incorporated into the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues,* 4th Edition classification system. Occasionally, a diagnosis may be based primarily on characteristic histologic features alone or in combination with clinical characteristics of the disease such as the presence or absence of a virus. Therefore, any combination of disease-specific characteristics may be described microscopically (histology/morphology), or may be identified by immunohistocytochemistry test, or identified by a specific immunophenotype or genetic abnormality. Part or all of these descriptive characteristics may be included in a new or updated hematopoietic or lymphoid neoplasm term or description (preferred term or synonym) or even in the disease classification (group) to which a specific disease entity may be assigned.

Several newly recognized conditions have been added to the 2008 classification. In addition, some conditions previously classified as borderline malignancy are now to be treated as malignant disease. The current classification divides hematopoietic and lymphoid neoplasms according to lineage. Three primary lines are used in the classification: myeloid, lymphoid, and histiocytic/dendritic. The 2008 *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues,* 4th Edition is used as the basis for this coding manual. The coding manual includes tables that describe the classification of disease along cell lines (lineage tables). Lineage tables are included in Appendix D.

The World *Health Organization Classification of Tumors of Hematopoietic and Lymphoid Tissues,* 3rd Edition, published in 2001, was based on principles defined in the Revised European-American Classification of Lymphoid Neoplasms (REAL), originally published by the International Lymphoma Study Group in 1994. Both the REAL and current classifications group borderline and malignant tumors into broad categories by hematologic lineage: myeloid, lymphoid, histiocytic/dendritic, and mast cell. Within these broad categories or phenotypes, tumors may present in solid or circulating phases. Solid phase is the presence of malignant cells in tissue, such as lymph nodes, soft tissues, or organs; generally these have historically been called lymphomas. The circulating phase is characterized by the presence of malignant cells in the circulating blood or bone marrow; historically these have been called leukemias. According to the introduction to the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues,* 4th Edition, the "…distinction between them (lymphomas and leukemias) is artificial. Thus B-cell chronic lymphocytic leukemia and B-cell small lymphocytic lymphoma are simply different manifestations of the same neoplasm, as are lymphoblastic lymphomas and lymphoblastic leukemias and Burkitt lymphoma and Burkitt leukemia" (2001 WHO Classification, page 13).

Although each of these pairs of diagnoses is histopathologically the same malignant cell with different presentations, they have different morphology code numbers in the *International Classification of Diseases for Oncology*, Third Edition (ICD-O-3). This is because ICD-O-3 is a subset of the *International Statistical Classification of Diseases and Related Health Problems*, Tenth Revision (ICD-10), in which the distinction between lymphomas and leukemias was maintained. ICD-10 was originally published in 1990, prior to the publication of the REAL classification that introduced the concept of grouping lymphoid and hematopoietic malignancies by phenotype rather than morphologic characteristics and clinical presentation. In order to ensure compatibility with ICD-10, the Third Edition of ICD-O differs from the structure of the WHO classification of hematologic malignancies.

The concept of cross-referencing two histology codes in ICD-O-3 was necessary because ICD-10 had not yet caught up with current medical concepts in the area of classification of lymphoma and leukemia. The following is noted in the introductory text of ICD-O-3 (page 14):

> ***Compatibility with ICD-10***
> In order to ensure compatibility with ICD-10, there are a number of ways in which the Third Edition of ICD-O differs from the structure of the WHO classification of hematologic malignancies. Separate codes have been allocated to B-cell chronic lymphocytic leukemia and B-cell small lymphocytic lymphoma. These are now recognized to be exactly the same entity, and for presentation of data these categories may therefore be combined. The same argument applies to lymphoblastic lymphoma and acute lymphoblastic leukemia, which are now regarded as the same disease but for which separate codes are provided.

The existence of dual codes for the same WHO classification entities is further discussed in the first errata for ICD-O-3 (5-22-2001):

> ***6. Assigning topography for hematopoietic diseases*** According to the medical understanding on which the World Health Organization Classification of Hematopoietic Neoplasms is based, some lymphomas and leukemias are the same disease with different presentations. For example, the WHO Classification lists B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (BCCLL/SLL) as a single entity, the same disease at different stages. The hemato-pathologists on the ICD-O-3 development committee recommended a single code number to represent the disease. However, since ICD-O is a subset of ICD-10 and ICD-10 is used to code mortality throughout the world, if a single ICD-O-3 code were used, there would be no way to determine whether a death was due to lymphoma or leukemia which are coded separately in ICD-10. As a result, it was necessary to retain separate codes for chronic lymphocytic leukemia and small lymphocytic lymphoma and link them. Thus, for the first time in ICD-O editions, some single disease entities are listed in two different categories and cross-referenced with the notation (see also M-9----). The topographic or primary site code for a diagnosis such as BCCLL/SLL depends on where the disease is diagnosed: if disease is diagnosed only in the blood or bone marrow, code the primary site to C42.1, bone marrow and assign the leukemia morphology code. For purposes of analysis according to the WHO Classification, cases from both morphology codes should be aggregated.

Resources used

World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues. 4th Edition, World Health Organization, 2008.
World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues. 3rd Edition, World Health Organization, 2001.
International Classification of Diseases for Oncology, Third Edition. World Health Organization, 2000.
Essential Haematology, Fifth edition. Hoffbrand AV, Moss PAH and Pettit JE. Blackwell Publishing, 2006.
Abstracting and Coding Guide for the Hematopoietic Diseases. National Cancer Institute, 2002.

Obsolete Terms as Defined in ICD-O-3
Hematopoietic and Lymphoid Neoplasms

The following tables identify terms that are no longer used to describe diseases and to display the current term used for that disease. The terms designated obsolete [obs] are based on ICD-O-3 term and category assignment only. Obsolete [obs] designations have not been updated to match the 2008 WHO Classification. Revised obsolete [obs] term designations matching the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition will be available when a revision or new edition of ICD-O is published either as an addendum to ICD-O-3 or as ICD-O-4. Note that the ICD-O-3 code does not change, only the name that is commonly used to describe the disease. The tables also provide information on the origin of the term and the date the term became obsolete.

( ) indicates an optional term in the phrase

Table A1: Histiocytic and Dendritic Cell Neoplasm Obsolete Terms

Obsolete Term	Notes	ICD-O-3 Code	Current Term
Histiocytic medullary reticulosis	*Term used as early as 1939; Obsolete as of 1987 with publication of Langerhans cell histiocytosis terminology*	9750/3	Malignant histiocytosis
Nonlipid reticuloendotheliosis	*Term used as early as 1955; Obsolete as of 1987 with publication of Langerhans cell histiocytosis terminology*	9754/3	Langerhans cell histiocytosis, disseminated

Table A2: Hodgkin Lymphoma (Hodgkin Disease) Obsolete Terms

Obsolete Term	Notes	ICD-O-3 Code	Current Term
Hodgkin disease, lymphocyte predominance, diffuse	*Source: Lukes-Butler classification, 1966; Obsolete: REAL classification 1994*	9651/3	Lymphocyte-rich Hodgkin lymphoma
Hodgkin disease, lymphocyte predominance, NOS	*Source : Rye classification, 1966 ; Obsolete : REAL classification 1994*	9651/3	Lymphocyte-rich Hodgkin lymphoma
Hodgkin disease, lymphocytic-histiocytic predominance	*Source: Lukes-Butler classification, 1966; Obsolete as of REAL classification 1994*	9651/3	Lymphocyte-rich Hodgkin lymphoma
Hodgkin nodular paragranuloma	*Source : Jackson-Parker Classification, 1944 ; Obsolete : 1966*	9659/3	Nodular lymphocyte-predominance Hodgkin lymphoma
Hodgkin paragranuloma, NOS	*Source : Jackson-Parker Classification, 1944 ; Obsolete : 1966*	9659/3	Nodular lymphocyte-predominance Hodgkin lymphoma

Table A3: Lymphoid Neoplasm Obsolete Terms

Classification	Obsolete Term	Current Term	ICD-O-3 Code
B-cell Neoplasms	Immunocytoma	Lymphoplasmacytic lymphoma	9671/3
	Plasmacytoid lymphoma		
	Plasmacytic lymphoma		
	Mantle zone lymphoma	Mantle cell lymphoma	9673/3
	Intermediate differentiation diffuse lymphocytic lymphoma		
	Centrocytic lymphoma		
	Histiocytic lymphoma, NOS	Diffuse large B-cell lymphoma	9680/3
	Large cell cleaved and noncleaved lymphoma		
	Large cell diffuse lymphoma, NOS		
	Large cleaved cell lymphoma, NOS		
	Large cell cleaved lymphoma, NOS		
	Noncleaved diffuse lymphoma, NOS		
	Burkitt tumor	Burkitt lymphoma	9687/3
	Undifferentiated lymphoma, Burkitt type		
	Small noncleaved lymphoma, Burkitt type		
	Acute leukemia, Burkitt type	Burkitt cell leukemia	9826/3
	B-ALL		
	FAB L3		
	Centroblastic-centrocytic follicular lymphoma	Follicular lymphoma	9690/3
	Nodular lymphoma, NOS		
	Nodular lymphocytic lymphoma, NOS		
	Mixed small cleaved and large cell follicular lymphoma	Follicular lymphoma, grade 2	9691/3
	Mixed lymphocytic-histiocytic nodular lymphoma		
	Mixed cell type follicular lymphoma		
	Mixed cell type nodular lymphoma		
	Small cleaved cell follicular lymphoma	Follicular lymphoma, grade 1	9695/3
	Lymphocytic poorly differentiated nodular lymphoma		
	Large cell noncleaved follicular lymphoma	Follicular lymphoma, grade 3A Follicular lymphoma, grade 3B	9698/3
	Histiocytic nodular lymphoma		
	Noncleaved cell follicular lymphoma, NOS		
	Large cleaved cell follicular lymphoma		
	Lymphocytic well differentiated nodular lymphoma		
T-Cell and NK-Cell Neoplasms	Angiocentric T-cell lymphoma	NK/T-cell lymphoma, nasal and nasal-type	9719/3
	Malignant reticulosis, NOS		
	Malignant midline reticulosis		
	Polymorphic reticulosis		

Classification	Obsolete Term	Current Term	ICD-O-3 Code
	Large cell (Ki-1 positive) lymphoma	Anaplastic large cell lymphoma, ALK positive	9714/3
	Peripheral T-cell lymphoma, AILD (Angioimmunoblastic Lymphadenopathy with Dysproteinemia)	Angioimmunoblastic T-cell lymphoma	9705/3
	Angioimmunoblastic lymphoma		

Table A4: Myeloid Neoplasm Obsolete Terms

Classification	Obsolete Term	Notes	Current Term	ICD-O-3 Code
Acute Myeloid Leukemias	Acute erythemia	*Listed as separate code in ICD-O-1; code changed to 9840 in ICD-O-3; Obsolete: as of FAB classification 1986*	Acute myeloid leukemia, M6 type	
	Di Guglielmo disease	*Eponym from as early as 1928; listed as synonym for acute erythemia in ICD-O-1 9840/3*	Acute erythroid leukemia	9840/3
	Acute erythremic myelosis	*Listed as synonym for acute erythemia in ICD-O-1*		
	Malignant myelosclerosis	*Term first used in 1963; Obsolete: as of FAB classification 1986*	Acute panmyelosis with myelofibrosis	9931/3
Chronic Myeloproliferative Diseases	Chronic erythemia	*Term used as early as 1892; not in ICD-O-1 or ICD-O-2; obsolete: 2001*	Polycythemia vera	9950/3
Myelodysplastic/ Myeloproliferative Diseases	Chronic myelomonocytic leukemia in transformation	*Source: French American British classification 1986; Obsolete: 2001*	Chronic myelomonocytic leukemia	9945/3
Myelodysplastic Syndromes	Preleukemia	*Term used as early as 1949; not in ICD-O-1; listed as synonym of MDS in ICD-O-2*	Myelodysplastic syndrome, NOS	9989/3
	Preleukemic syndrome	*Term first used in 1973; not in ICD-O-1; listed as synonym of MDS in ICD-O-2*		

REFERENCES
1956 Publication of Rappaport classification of non-Hodgkin lymphomas
1966 Publication of Rye classification of Hodgkin lymphomas
1982 Publication of Working Formulation
1986 Publication of revised FAB classification (variously reported as 1982, 1985, or 1986)
2001 Publication of WHO classification, 3rd ed., and implementation of ICD-O-3
UICC TNM Supplement, 3rd ed., Wittekind, Greene, Henson, Hutter, Sobin

Appendix B
WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues
Histology Lineage

Note: Only the WHO Preferred Terms are listed.
Use the Hematopoietic Database to identify synonyms that correspond to the WHO Preferred Term.

Table B1: Myeloproliferative Neoplasms

WHO Preferred Term	ICD-O- 3
Chronic eosinophilic leukemia, NOS	9964/3
Chronic myelogenous leukemia, BCR-ABL1 positive	9875/3
Chronic neutrophilic leukemia	9963/3
Cutaneous mastocytosis	9740/1
Essential thrombocythemia	9962/3
Mast cell leukemia	9742/3
Mast cell sarcoma	9740/3
Myeloproliferative neoplasm, unclassifiable	9975/3
Polycythemia vera	9950/3
Primary myelofibrosis	9961/3
Systemic mastocytosis	9741/3

Table B2: Myeloid and Lymphoid Neoplasms with Eosinophilia and Abnormalities of PDGFRA, PDGFRB or FGFR1

WHO Preferred Term	ICD-O-3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Myeloid and lymphoid neoplasm with PDGFRA rearrangement	9965/3
Myeloid neoplasm with PDGFRB rearrangement	9966/3

Table B3: Myelodysplastic / Myeloproliferative Neoplasms

WHO Preferred Term	ICD-O-3
Atypical chronic myeloid leukemia, BCR-ABL1 negative	9876/3
Chronic myelomonocytic leukemia	9945/3
Juvenile myelomonocytic leukemia	9946/3
Myelodysplastic/myeloproliferative neoplasm, unclassifiable	9975/3
Refractory anemia with ring sideroblasts	9982/3

Table B4: Myelodysplastic Syndromes

WHO Preferred Term	ICD-O-3
Myelodysplastic syndrome associated with isolated del(5q)	9986/3
Myelodysplastic syndrome, unclassifiable	9989/3
Refractory anemia	9980/3
Refractory anemia with excess blasts	9983/3
Refractory anemia with ring sideroblasts	9982/3
Refractory cytopenia of childhood	9985/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3

Table B5: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms

WHO Preferred Term	ICD-O-3
Acute myeloid leukemias with recurrent genetic abnormalities	
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFB-MYH11	9871/3
Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26;2); RPN1-EVI1	9869/3
Acute myeloid leukemia with t(8;21)t(q22;q22); RUNX1-RUNX1T1	9896/3
Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL	9897/3
Acute promyelocytic leukemia (AML with t(15;17)(q22;q12), PML/RARA	9866/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3
Acute myeloid leukemia with myelodysplasia-related changes	9895/3
Therapy-related myeloid neoplasm	9920/3
Acute myeloid leukemia, NOS	9861/3
Acute monoblastic and monocytic leukemia	9891/3
Acute myeloid leukemia with minimal differentiation	9872/3
Acute myeloid leukemia without maturation	9873/3
Acute myeloid leukemia with maturation	9874/3
Acute myelomonocytic leukemia	9867/3
Acute erythroid leukemia	9840/3
Acute megakaryoblastic leukemia	9910/3
Acute basophilic leukemia	9870/3
Acute panmyelosis with myelofibrosis	9931/3
Myeloid sarcoma	9930/3
Myeloid proliferations related to Down syndrome	*No Code*
Transient abnormal myelopoiesis	9898/1

WHO Preferred Term	ICD-O-3
Myeloid leukemia associated with Down syndrome	9898/3
Blastic plasmacytoid dendritic cell neoplasm	9727/3

Table B6: Acute Leukemia of Ambiguous Lineage

WHO Preferred Term	ICD-O-3
Acute undifferentiated leukemia	9801/3
Mixed phenotype acute leukemia with t(v;11q2); MLL rearranged	9807/3
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Natural killer (NK) cell lymphoblastic leukemia/lymphoma	*No Code*

Table B7: Precursor Lymphoid Neoplasms

WHO Preferred Term	ICD-O-3
B lymphoblastic leukemia/lymphoma	No Code
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with recurrent genetic abnormalities	No Code
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A-PBX1 (TCF3-PBX1)	9818/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3
T lymphoblastic leukemia/lymphoma	9837/3

Table B8: Mature B-Cell Neoplasms

WHO Preferred Term	ICD-O-3
ALK positive large B-cell lymphoma	9737/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma	9596/3
B-cell prolymphocytic leukemia	9833/3
Burkitt lymphoma	9687/3
Chronic lymphocytic leukemia/small lymphocytic lymphoma	9823/3
Diffuse large B-cell lymphoma (DLBCL), NOS	9680/3
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)	9699/3
Extraosseous plasmacytoma	9734/3
Follicular lymphoma	9690/3

WHO Preferred Term	ICD-O-3
Hairy cell leukemia	9940/3
Heavy chain disease	9762/3
Intravascular large B-cell lymphoma	9712/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Lymphomatoid granulomatosis	9766/1
Lymphoplasmacytic lymphoma	9671/3
Mantle cell lymphoma	9673/3
Plasma cell myeloma	9732/3
Plasmablastic lymphoma	9735/3
Primary cutaneous follicle centre lymphoma	9597/3
Primary effusion lymphoma	9678/3
Primary mediastinal (thymic) large B-cell lymphoma	9679/3
Solitary plasmacytoma of bone	9731/3
Splenic B-cell lymphoma/leukemia, unclassifiable	9591/3
Splenic marginal zone lymphoma	9689/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3
Waldenstrom Macroglobulinemia	9761/3

Table B9: Mature T-Cell and NK-Cell Neoplasms

WHO Preferred Term	ICD-O-3
Adult T-cell leukemia/lymphoma	9827/3
Aggressive NK-cell leukemia	9948/3
Anaplastic large cell lymphoma, ALK positive	9714/3
Angioimmunoblastic T-cell lymphoma	9705/3
Chronic lymphoproliferative disorder of NK-cells	9831/3
Enteropathy-associated T-cell lymphoma	9717/3
Extranodal NK/T cell lymphoma, nasal type	9719/3
Hepatosplenic T-cell lymphoma	9716/3
Hydroa vacciniforme-like lymphoma	9725/3
Lymphomatoid papulosis	9718/1
Mycosis fungoides	9700/3
Peripheral T-cell lymphoma, NOS	9702/3
Primary cutaneous anaplastic large cell lymphoma	9718/3
Primary cutaneous CD30 positive T-cell lymphoproliferative disorders	*No Code*
Primary cutaneous CD4 positive small/medium cell T-cell lymphoma	9709/3
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Sezary syndrome	9701/3
Subcutaneous panniculitis-like T-cell lymphoma	9708/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
T-cell prolymphocytic leukemia	9834/3

Table B10: Hodgkin Lymphoma

WHO Preferred Term	ICD-O-3
Classical Hodgkin lymphoma	9650/3
Lymphocyte-depleted classical Hodgkin lymphoma	9653/3
Lymphocyte-rich classical Hodgkin lymphoma	9651/3
Mixed cellularity classical Hodgkin lymphoma	9652/3
Nodular lymphocyte predominant Hodgkin lymphoma	9659/3
Nodular sclerosis classical Hodgkin lymphoma	9663/3

Table B11: Histiocytic and Dendritic Cell Neoplasms

WHO Preferred Term	ICD-O-3
Disseminated juvenile xsanthogranuloma	No Code
Fibroblastic reticular cell tumor	9759/3
Follicular dendritic cell sarcoma	9758/3
Histiocytic sarcoma	9755/3
Indeterminate dendritic cell tumor	9757/3
Langerhans cell histiocytosis	9751/3
Langerhans cell sarcoma	9756/3

Table B12: Post-Transplant Lymphoproliferative Disorders (PTLD)

WHO Preferred Term	ICD-O-3
Early lesions	*No Code*
Classical Hodgkin lymphoma type PTLD	*
Infectious mononucleosis-like PTLD	9971/1
Monomorphic PTLD (B- and T/NK-cell types)	*
Plasmacytic hyperplasia	9971/1
Polymorphic PTLD	9971/3

*These lesions are classified according to the leukemia or lymphoma to which they correspond, and are assigned the respective ICD-O code.

Appendix C
Lymph Node/Lymph Node Chain Reference Table

Use this table with the Primary Site and Histology Rules to determine whether involved lymph nodes are in a single ICD-O-3 lymph node region or in multiple ICD-O-3 lymph node regions.

This table contains the names of lymph nodes that have the capsule and sinus structure of true lymph nodes. Lymphoid tissue such as that in the GI tract, tonsils, etc., is not represented in this table.

Note: Pathology reports may identify lymph nodes within most organs, the most common being breast, parotid gland, lung, and pancreas. The lymph nodes in these organs are called intra-(organ name) lymph nodes such as intramammary lymph nodes. We have included the most common intra-organ lymph nodes on this table. For an intra-organ lymph node not listed on the table, code to the ICD-O-3 topography code for that organ's regional lymph node chain(s).

Table C1: Lymph Node/Lymph Node Chain Reference Table

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Abdominal	C772	Intra-abdominal	Mesenteric
Anorectal	C775	Pelvic	Pelvic, right and left*
Anterior axillary	C773	Axilla or arm	Axillary, right and left*
Anterior cecal	C772	Intra-abdominal	Mesenteric
Anterior deep cervical	C770	Head, face and neck	Cervical, right and left*
Anterior jugular	C770	Head, face and neck	Cervical, right and left*
Aortic NOS; ascending aortic lateral aortic; lumbar aortic; para-aortic; peri-aortic	C772	Intra-abdominal	Para-aortic
Aortico-pulmonary window (subaortic)	C772	Intra-abdominal	Para-aortic
Appendiceal	C772	Intra-abdominal	Mesenteric
Ascending aortic	C772	Intra-abdominal	Para-aortic
Aselli's glands (nodes near pancreas)	C772	Intra-abdominal	Para-aortic
Auricular NOS; infra-auricular; pre-auricular; post-auricular; retro-auricular	C770	Head, face and neck	Cervical, right and left*
Axillary, lateral;	C773	Axilla or arm	Axillary, right and left*
Axillary; anterior	C773	Axilla or arm	Axillary, right and left*
Azygos (lower paratracheal)	C771	Intrathoracic	Mediastinal
Brachial	C773	Axilla or arm	Axillary, right and left*
Bronchial,; bronchopulmonary; hilar; proximal lobar; pulmonary root	C771	Intrathoracic	Hilar
Bronchopulmonary	C771	Intrathoracic	Hilar
Bronchopulmonary; bronchial hilar; proximal lobar; pulmonary root	C771	Intrathoracic	Hilar
Buccal	C770	Head, face and neck	Cervical, right and left*
Buccinator (facial)	C770	Head, face and neck	Cervical, right and left*

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Calot's node (cysto-hepatic triangle or hepato-biliary triangle)	C772	Intra-abdominal	Para-aortic
Cardiac	C771	Intrathoracic	Mediastinal
Cardial	C771	Intrathoracic	Mediastinal
Cardioesophageal	C771	Intrathoracic	Mediastinal
Carinal; tracheal bifurcation; tracheobronchial	C771	Intrathoracic	Mediastinal
Caval (para-)	C772	Intra-abdominal	Para-aortic
Cecal; anterior cecal: posterior cecal: prececal; retrocecal, NOS	C772	Intra-abdominal	Mesenteric
Celiac	C772	Intra-abdominal	Para-aortic
Central compartment (paralaryngeal, prelaryngeal [Delphian]) adjacent to thyroid gland.	C770	Head, face and neck	Cervical, right and left*
Cervical NOS, anterior deep cervical; deep cervical (scalene); lower deep cervical; upper/superior cervical; lower/inferior; middle deep cervical; posterior cervical (spinal accessory); transverse cervical (supraclavicular)	C770	Head, face and neck	Cervical, right and left*
Cloquet's node (inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Colic NOS, ileocolic, mesocolic, middle (right)	C772	Intra-abdominal	Mesenteric
Common bile duct	C772	Intra-abdominal	Para-aortic
Cubital	C773	Axilla or arm	Axillary, right and left*
Cystic duct	C772	Intra-abdominal	Para-aortic
Deep cervical (laterotracheal)	C771	Intrathoracic	Cervical, right and left*
Deep inguinal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Delphian node (prepharyngeal), adjacent to thyroid gland.	C770	Head, face and neck	Cervical, right and left*
Deltopectoral	C773	Axilla or arm	Axillary, right and left*
Diaphragmatic, sub	C771	Intrathoracic	Mediastinal
Duodenal	C772	Intra-abdominal	Para-aortic
Epicolic (Foramen of Winslow, omental)	C772	Intra-abdominal	Mesenteric
Epitrochlear	C773	Axilla or arm	Axillary, right and left*
Esophageal (para-, peri-)	C771	Intrathoracic	Mediastinal
Facial	C770	Head, face and neck	Cervical, right and left*
Femoral (superficial inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Foramen of Winslow (epicolic, omental)	C772	Intra-abdominal	Mesenteric
Gastric NOS, left (superior), gastrocolic; right (inferior gastric);	C772	Intra-abdominal	Mesenteric
Gastrocolic (Gastric)	C772	Intra-abdominal	Mesenteric
Gastroduodenal	C772	Intra-abdominal	Mesenteric
Gastroepiploic (gastro-omental)	C772	Intra-abdominal	Mesenteric
Gastrohepatic	C772	Intra-abdominal	Mesenteric
Gastro-omental (gastroepiploic)	C772	Intra-abdominal	Mesenteric
Gastropancreatic	C772	Intra-abdominal	Mesenteric
Gerota's node (promontorial, middle sacral)	C775	Pelvic	Para-aortic

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Greater curvature	C772	Intra-abdominal	Mesenteric
Greater omental	C772	Intra-abdominal	Mesenteric
Groin	C774	Inguinal region or leg	Inguino-femoral, right and left*
Hemorrhoidal NOS; inferior; middle; superior	C775	Pelvic	Pelvic, right and left*
Hepatic, NOS; hepatic artery; hepatic pedicle; hepatic, inferior vena cava; porta hepatis (hilar)	C772	Intra-abdominal	Para-aortic
Hepatoduodenal ligament	C772	Intra-abdominal	Para-aortic
Hilar (splenic)	C772	Intra-abdominal	Mesenteric
Hilar [in hilus of liver] (porta hepatis, portal)	C772	Intra-abdominal	Para-aortic
Hilar, bronchial	C771	Intrathoracic	Hilar
Hilar, hepatoduodenal ligament	C772	Intra-abdominal	Para-aortic
Hilar, pulmonary root	C771	Intrathoracic	Hilar
Hilar; bronchopulmonary, proximal lobar, pulmonary root	C771	Intrathoracic	Hilar
Hypogastric (internal iliac)	C775	Pelvic	Pelvic, right and left*
Ileocolic	C772	Intra-abdominal	Mesenteric
Iliac, common	C775	Pelvic	Pelvic, right and left*
Iliac, external	C775	Pelvic	Pelvic, right and left*
Iliac, internal (hypogastric, obturator)	C775	Pelvic	Pelvic, right and left*
Inferior deep cervical (scalene)	C770	Head, face and neck	Cervical, right and left*
Inferior deep jugular	C770	Head, face and neck	Cervical, right and left*
Inferior gastric (right gastric)	C772	Intra-abdominal	Mesenteric
Inferior hemhorrhoidal	C775	Pelvic	Pelvic, right and left*
Inferior vena cava	C772	Intra-abdominal	Para-aortic
Infra-auricular	C770	Head, face and neck	Cervical, right and left*
Infraclavicular (subclavicular)	C773	Axilla or arm	Infraclavicular, right and left*
Infrapyloric (subpyloric)	C772	Intra-abdominal	Para-aortic
Inguinal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Inguinal NOS; deep, superficial (subinguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Innominate (thoracic)	C771	Intrathoracic	Mediastinal
Interaortocaval	C772	Intra-abdominal	Para-aortic
Intercostal	C771	Intrathoracic	Mediastinal
Interlobar (within the lung)/intrapulmonary	C771	Intrathoracic	Mediastinal
Internal iliac (hemorrhoidal)	C775	Pelvic	Pelvic, right and left*
Internal jugular	C770	Head, face and neck	Cervical, right and left*
Internal mammary (parasternal)	C771	Intrathoracic	Mediastinal
Interpectoral	C773	Axilla or arm	Axillary, right and left*
Intestinal	C772	Intra-abdominal	Mesenteric
Intra-abdominal	C772	Intra-abdominal	Mesenteric
Intrabronchial, NOS	C771	Intrathoracic	Hilar

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Intramammary lymph node	C773	Axilla or arm	Axillary, right and left*
Intrapancreatic lymph node	C772	Intra-abdominal	Para-aortic
Intraparotid	C770	Head, face and neck	Cervical, right and left*
Intrapelvic	C775	Pelvic	Pelvic, right and left*
Intrapulmonary (within the lung)	C771	Intrathoracic	Mediastinal
Intrapulmonary, segmental/subsegmental	C771	Intrathoracic	Mediastinal
Jugular, lower, mid, upper, internal	C770	Head, face and neck	Cervical, right and left*
Jugulodigastric (subdigastric)	C770	Head, face and neck	Cervical, right and left*
Jugulo-omohyoid (supraomohyoid)	C770	Head, face and neck	Cervical, right and left*
Lateral aortic (lumbar)	C772	Intra-abdominal	Para-aortic
Lateral compartment (jugular, mid and lower; supraclavicular; upper deep jugular; spinal accessory; retropharyngeal; submandibular; submental)l	C770	Head, face and neck	Cervical, right and left*
Lateral jugular	C770	Head, face and neck	Cervical, right and left*
Laterotracheal (anterior deep cervical)	C771	Intrathoracic	Cervical, right and left*
Left (superior) gastrocolic	C772	Intra-abdominal	Mesenteric
Leg/Lower limb	C774	Inguinal region or leg	Inguino-femoral, right and left*
Lesser curvature	C772	Intra-abdominal	Mesenteric
Lesser omental	C772	Intra-abdominal	Mesenteric
Lineal (splenic)	C772	Intra-abdominal	Mesenteric
Lobar, proximal (pulmonary)	C771	Intrathoracic	Hilar
Lobar/intrapulmonary	C771	Intrathoracic	Hilar
Lower jugular	C770	Head, face and neck	Cervical, right and left*
Lower peratracheal	C771	Intrathoracic	Mediastinal
Lower periesophageal (intrathoracic esophagus)	C771	Intrathoracic	Mediastinal
Lower peritracheal	C771	Intrathoracic	Mediastinal
Lumbar	C771	Intra-abdominal	Pelvis, right and left*
Lumbar aortic	C772	Intra-abdominal	Para-aortic
Mandibular	C770	Head, face and neck	Cervical, right and left*
Mastoid (postauricular, retro-auricular)	C770	Head, face and neck	Cervical, right and left*
Mediastinal, anterior	C771	Intrathoracic	Mediastinal
Mediastinal, NOS	C771	Intrathoracic	Mediastinal
Mediastinal, posterior (tracheoesophageal)	C771	Intrathoracic	Mediastinal
Mediastinal, superior	C771	Intrathoracic	Mediastinal
Mesenteric	C772	Intra-abdominal	Mesenteric
Mesenteric, inferior	C772	Intra-abdominal	Mesenteric
Mesenteric, sigmoid	C772	Intra-abdominal	Mesenteric
Mesenteric, superior	C772	Intra-abdominal	Mesenteric

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Mesocolic	C772	Intra-abdominal	Mesenteric
Mid jugular	C770	Head, face and neck	Cervical, right and left*
Midcolic	C772	Intra-abdominal	Pelvic, right and left*
Middle (right) colic	C772	Intra-abdominal	Mesenteric
Middle hemhorrhoidal	C775	Pelvic	Pelvic, right and left*
Middle sacral	C775	Pelvic	Pelvic, right and left*
Nasolabial	C770	Head, face and neck	Cervical, right and left*
Node of Cloquet's or Rosenmuller (highest deep inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Obturator	C775	Pelvic	Pelvic, right and left*
Occipital; suboccipital	C770	Head, face and neck	Cervical, right and left*
Omental	C772	Intra-abdominal	Mesenteric
Pancreatic; Aselli's glands (nodes near pancreas); parapancreatic; peripancreatic	C772	Intra-abdominal	Para-aortic
Pancreaticoduodenal	C772	Intra-abdominal	Para-aortic
Pancreaticosplenic (pancreaticolineal)	C772	Intra-abdominal	Mesenteric
Para-aortic	C772	Intra-abdominal	Para-aortic
Parabronchial	C771	Intrathoracic	Mediastinal
Paracardial	C772	Intra-abdominal	Mesenteric
Paracaval	C772	Intra-abdominal	Para-aortic
Paracervical	C775	Pelvic	Pelvic, right and left*
Paracolic/pericolic	C772	Intra-abdominal	Para-aortic
Paraesophageal	C771	Intrathoracic	Mediastinal
Paralaryngeal	C770	Head, face and neck	Cervical, right and left*
Parametrial	C775	Pelvic	Pelvic, right and left*
Parapancreatic	C772	Intra-abdominal	Para-aortic
Parapharyngeal	C770	Head, face and neck	Cervical, right and left*
Parasternal (internal mammary)	C771	Intrathoracic	Mediastinal
Paratracheal	C771	Intrathoracic	Mediastinal
Paratracheal, lower	C771	Intrathoracic	Mediastinal
Parotid (peri-)	C770	Head, face and neck	Cervical, right and left*
Pectoral (anterior axillary)	C773	Axilla or arm	Axillary, right and left*
Pelvic, NOS	C775	Pelvic	Pelvic, right and left*
Peratracheal, lower	C771	Intrathoracic	Mediastinal
Peri-aortic	C772	Intra-abdominal	Para-aortic
Peribronchial; parabronchial	C771	Intrathoracic	Mediastinal
Pericardial	C771	Intrathoracic	Mediastinal
Pericaval	C772	Intra-abdominal	Para-aortic
Pericholedochal	C772	Intra-abdominal	Para-aortic

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Pericolic	C772	Intra-abdominal	Mesenteric
Periduodenal	C772	Intra-abdominal	Para-aortic
Periesophageal	C771	Intrathoracic	Mediastinal
Perigastric, except cardiac	C772	Intra-abdominal	Mesenteric
Peripancreatic	C772	Intra-abdominal	Para-aortic
Periparotid	C770	Head, face and neck	Cervical, right and left*
Periportal	C772	Intra-abdominal	Pelvic, right and left*
Periprostatic	C775	Pelvic	Pelvic, right and left*
Perirectal	C775	Pelvic	Pelvic, right and left*
Peritracheal, lower	C771	Intrathoracic	Mediastinal
Periureteral	C772	Intra-abdominal	Para-aortic
Perivesical	C775	Pelvic	Pelvic, right and left*
Pharyngeal; Delphian node; prepharyngeal; retropharyngeal	C770	Head, face and neck	Cervical, right and left*
Phrenic; inferior phrenic vein; superior phrenic vein	C771	Intra-thoracic	Mediastinal
Popliteal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Porta hepatis [in hilus of liver]	C772	Intra-abdominal	Para-aortic
Portal (portal vein)	C772	Intra-abdominal	Para-aortic
Postauricular	C770	Head, face and neck	Cervical, right and left*
Posterior axillary	C773	Axilla or arm	Axillary, right and left*
Posterior cecal	C772	Intra-abdominal	Para-aortic
Posterior mediastinal	C771	Intrathoracic	Mediastinal
Posterior triangle (spinal accessory and transverse cervical)	C770	Head, face and neck	Cervical, right and left*
Preaortic	C772	Intra-abdominal	Para-aortic
Pre-auricular	C770	Head, face and neck	Cervical, right and left*
Precarinal	C771	Intrathoracic	Mediastinal
Prececal	C772	Intra-abdominal	Mesenteric
Prelaryngeal	C770	Head, face and neck	Cervical, right and left*
Prepharyngeal	C770	Head, face and neck	Cervical, right and left*
Presymphsial	C775	Pelvic	Pelvic, right and left*
Pretracheal	C770	Head, face and neck	Cervical, right and left*
Promontorial	C775	Pelvic	Pelvic, right and left*
Proximal lobar	C771	Intrathoracic	Hilar
Proximal mesentery	C772	Intra-abdominal	Mesenteric
Pulmonary ligament	C771	Intrathoracic	Mediastinal
Pulmonary root	C771	Intrathoracic	Hilar
Pulmonary, NOS	C771	Intrathoracic	Hilar
Pyloric, Infra (subpyloric)	C772	Intra-abdominal	Para-aortic
Pyloric; suprapyloric	C772	Intra-abdominal	Para-aortic

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Rectal	C775	Pelvic	Pelvic, right and left*
Recurrent laryngeal (laterotracheal, anterior deep cervical)	C770	Head, face and neck	Cervical, right and left*
Recurrent pharyngeal	C770	Head, face and neck	Cervical, right and left*
Renal hilar	C772	Intra-abdominal	Para-aortic
Retroaortic	C772	Intra-abdominal	Para-aortic
Retroauricular (mastoid)	C770	Head, face and neck	Cervical, right and left*
Retrocaval	C772	Intra-abdominal	Para-aortic
Retrocecal	C772	Intra-abdominal	Para-aortic
Retrocecal (posterior cecal)	C772	Intra-abdominal	Para-aortic
Retrocrural	C771	Intra-thoracic	Mediastinal
Retroperitoneal	C772	Intra-abdominal	Para-aortic
Retropharyngeal	C770	Head, face and neck	Cervical, right and left*
Retrotracheal	C771	Intrathoracic	Mediastinal
Right (inferior) gastric	C772	Intra-abdominal	Mesenteric
Rosenmuller's node (inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Rosenmuller's-Cloquet's nodes (inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Rotter's nodes (interpectoral between major and minor pectoralis)	C773	Axilla or arm	Axillary, right and left*
Rouviere's node (retropharyngeal)	C770	Head, face and neck	Cervical, right and left*
Sacral, lateral (laterosacral)	C775	Pelvic	Pelvic, right and left*
Sacral, middle (promontorial/promontory) (Gerota's node)	C775	Pelvic	Pelvic, right and left*
Sacral, NOS	C775	Pelvic	Pelvic, right and left*
Sacral, presacral	C775	Pelvic	Pelvic, right and left*
Sacral, uterosacral	C774	Pelvic	Pelvic, right and left*
Scalene (inferior deep cervical)	C770	Head, face and neck	Cervical, right and left*
Sigmoidal (sigmoid mesenteric)	C772	Intra-abdominal	Mesenteric
Spermatic vein	C774	Inguinal region or leg	Inguino-femoral, right and left*
Spinal accessory (posterior cervical)	C770	Head, face and neck	Cervical, right and left*
Splenic (hilar)	C772	Intra-abdominal	Mesenteric
Splenic (lienal)	C772	Intra-abdominal	Mesenteric
Subaortic (aortico-pulmonary window)	C772	Intra-abdominal	Mediastinal
Subcarinal	C771	Intrathoracic	Mediastinal
Subclavian (apical)	C770	Head, face and neck	Cervical, right and left*
Subclavicular (infraclavicular)	C773	Axilla or arm	Infraclavicular, right and left*
Subdigastric	C770	Head, face and neck	Cervical, right and left*
Subinguinal (superficial inguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Sublingual	C770	Head, face and neck	Cervical, right and left*
Submandibular (submaxillary)	C770	Head, face and neck	Cervical, right and left*
Submental	C770	Head, face and neck	Cervical, right and left*

Lymph Node/Lymph Node Chain	ICD-O-3 Code	ICD-O-3 Lymph Node Region(s)	AJCC Lymph Node Region(s)
Suboccipital	C770	Head, face and neck	Cervical, right and left*
Subpleural (in the periphery of the lung)	C771	Intrathoracic	Mediastinal
Subpyloric	C772	Intra-abdominal	Para-aortic
Subscapular (posterior axillary)	C773	Axilla or arm	Axillary, right and left*
Superficial inguinal (subinguinal)	C774	Inguinal region or leg	Inguino-femoral, right and left*
Superior gastrocolic (left gastrocolic)	C772	Intra-abdominal	Mesenteric
Superior hemhorrhoidal	C775	Pelvic	Pelvic, right and left*
Superior hilum	C772	Intra-abdominal	Pelvic, right and left*
Superior jugular	C770	Head, face and neck	Cervical, right and left*
Superior mesenteric	C772	Intra-abdominal	Pelvic, right and left*
Superior rectal	C775	Pelvic	Pelvic, right and left*
Supraclavicular (transverse cervical)	C770	Head, face and neck	Cervical, right and left*
Supraomohyoid	C770	Head, face and neck	Cervical, right and left*
Suprapancreatic	C772	Intra-abdominal	Para-aortic
Suprapyloric	C772	Intra-abdominal	Para-aortic
Thoracic (innominate)	C771	Intrathoracic	Mediastinal
Thyroid	C770	Head, face and neck	Cervical, right and left*
Tibial	C774	Inguinal region or leg	Inguino-femoral, right and left*
Tracheal bifurcation	C771	Intrathoracic	Mediastinal
Tracheal; pretracheal; retrotracheal	C771	Intrathoracic	Mediastinal
Tracheobronchial	C771	Intrathoracic	Mediastinal
Tracheoesophageal (posterior mediastinal)	C771	Intrathoracic	Mediastinal
Transverse cervical (supraclavicular)	C770	Head, face and neck	Cervical, right and left*
Transverse cervical; posterior triangle; spinal accessory	C770	Head, face and neck	Cervical, right and left*
Trosier's node (left supraclavicular)	C770	Head, face, and neck	Cervical, right and left*
Upper jugular	C770	Head, face and neck	Cervical, right and left*
Virchow's node (supraclavicular)	C770	Head, face, and neck	Cervical, right and left*

*The right and left are separate regions per AJCC

Appendix D
New Histology Terms and Codes Hematopoietic and Lymphoid Neoplasms

Table D1a: New Histology Terms and Codes – Alphabetic List

Table D1a contains an alphabetic list of hematopoietic and lymphoid neoplasm histology codes and terms documented in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Ed. published in 2008. Use this table to code the histology when any of these more specific terms are the diagnosis. Column 1 is the more specific histology term; column 2 is the new code WHO proposed for that specific histology. These neoplasms are not newly reportable; they are more specific terms for diseases that would otherwise be coded in NOS categories. Do not use these codes for neoplasms diagnosed prior to 2010. The new codes will go into effect with cases diagnosed 1/1/2010 and after. There are no plans or mandates to identify 2008 and 2009 cases to recode using these codes.

New Histology Term	ICD-O Code
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); RPN1EVI1	9869/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3
ALK positive large B-cell lymphoma	9737/3
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A PBX1 (TCF3 PBX1)	9818/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3
Fibroblastic reticular cell tumor	9759/3
Hydroa vacciniforme-like lymphoma	9725/3
Intravascular large B-cell lymphoma	9712/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged	9807/3
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Myeloid and lymphoid neoplasms with PDGFRB rearrangement	9966/3
Myeloid and lymphoid neoplasms with PDGFRA rearrangement	9965/3
Myeloid leukemia associated with Down Syndrome	9898/3
Plasmablastic lymphoma	9735/3
Polymorphic PTLD	9971/3
Primary cutaneous follicle centre lymphoma	9597/3

New Histology Term	ICD-O Code
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
T lymphoblastic leukemia/lymphoma	9837/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3

Table D1b: New Histology Terms and Codes – Numeric List

Table D1b contains a numeric list of hematopoietic and lymphoid neoplasm histology codes and terms documented in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Ed. published in 2008. Use this table to code the histology when any of these more specific terms are the diagnosis. Column 1 is the more specific histology term; column 2 is the new code WHO proposed for that specific histology. These neoplasms are not newly reportable; they are more specific terms for diseases that would otherwise be coded in NOS categories. Do not use these codes for neoplasms diagnosed prior to 2010. The new codes will go into effect with cases diagnosed 1/1/2010 and after. There are no plans or mandates to identify 2008 and 2009 cases to recode using these codes.

New Histology Term	ICD-O Code
Primary cutaneous follicle centre lymphoma	9597/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3
Intravascular large B-cell lymphoma	9712/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
Hydroa vacciniforme-like lymphoma	9725/3
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Plasmablastic lymphoma	9735/3
ALK positive large B-cell lymphoma	9737/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Fibroblastic reticular cell tumor	9759/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged	9807/3
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A PBX1 (TCF3 PBX1)	9818/3
T lymphoblastic leukemia/lymphoma	9837/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3

New Histology Term	ICD-O Code
Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); RPN1EVI1	9869/3
Myeloid leukemia associated with Down Syndrome	9898/3
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Myeloid and lymphoid neoplasms with PDGFRA rearrangement	9965/3
Myeloid and lymphoid neoplasms with PDGFRB rearrangement	9966/3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Polymorphic PTLD	9971/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3

Table D2: Histologic Terms and Codes with Changes in Case Reportability (Newly Reportable Conditions)
Table D2 contains hematopoietic and lymphoid neoplasms with changes in behavior from /1 (borderline malignancy) to /3 (malignant). The changes in histology codes and terms are documented in the the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition published in 2008. Reporting these neoplasms will go into effect with cases diagnosed 1/1/2010 and after. There are no plans or mandates to collect 2008 and 2009 cases having these diagnoses.

Histology Term	ICD-O Code
Langerhans cell histiocytosis, NOS	9751/3
T-cell large granular lymphocytic leukemia / Chronic lymphoproliferative disorder of NK-cells	9831/3
Myeloproliferative neoplasm, unclassifiable / Myelodysplastic/Myeloproliferative neoplasm, unclassifiable	9975/3

Appendix E
Histology "NOS" Tables

Table E1: NOS Terms and Codes Alphabetical List

Use this table for PH Module 8.
Appendix E contains two tables that list "NOS" terms and histology codes that are non-specific terms for a group of neoplasms with more specific subtypes. Leukemia, NOS is a non-specific term for a group of neoplasms that includes all leukemias; acute, chronic, and any lineage.

This table is used with two rules.
Rule PH 38: When a patient has a prior history of any "NOS" and presents with a more specific neoplasm, do not automatically create a new abstract/primary. Check the Hematopoietic DB and Appendix B to see if the "NOS" and specific neoplasms are the "same primary." If so, follow the instructions in Rule PH 38.

Rule PH39: When a patient is diagnosed with any of these "NOS" terms and two or more specific histologies, check the Hematopoietic DB and Appendix B to see if the "NOS" and all of the specific histologies are the "same primary." If they are the same primary, this is a provisional diagnosis. In the absence of more information, follow the instructions in Rule PH39.

Note: ICD-O-3 uses the term "NOS" to designate those histologies that are not well-defined. Some have subtypes, others do not. Appendix E lists only those "NOS" terms that have subtypes and should be carefully checked to determine whether or not the current neoplasm is an additional primary or a more specific subtype.

Term	Code
Acute leukemia, NOS	9801/3
Acute myeloid leukemia, NOS	9861/3
Chronic myeloproliferative disease, NOS	9960/3
Follicular lymphoma, NOS	9690/3
Hodgkin lymphoma, NOS	9650/3
Leukemia, NOS	9800/3
Malignant lymphoma, non-Hodgkin, NOS	9591/3
Malignant lymphoma, NOS	9590/3
Myelodysplastic syndrome, NOS	9989/3
Myeloid leukemia, NOS	9860/3
Myeloproliferative disease, NOS	9975/3

Table E2: NOS Terms and Codes Numeric List

Use this table for PH Module 8.
Appendix E contains two tables that list "NOS" terms and histology codes that are non-specific terms for a group of neoplasms with more specific subtypes. Leukemia, NOS is a non-specific term for a group of neoplasms that includes all leukemias; acute, chronic, and any lineage.

This table is used with two rules.
Rule PH 38: When a patient has a prior history of any "NOS" and presents with a more specific neoplasm, do not automatically create a new abstract/primary. Check the Hematopoietic DB and Appendix B to see if the "NOS" and specific neoplasms are the "same primary." If so, follow the instructions in Rule PH 38.

Rule PH39: When a patient is diagnosed with any of these "NOS" terms and two or more specific histologies, check the Hematopoietic DB and Appendix B to see if the "NOS" and all of the specific histologies are the "same primary." If they are the same primary, this is a provisional diagnosis. In the absence of more information, follow the instructions in Rule PH39.

Note: ICD-O-3 uses the term "NOS" to designate those histologies that are not well-defined. Some have subtypes, others do not. Appendix E lists only those "NOS" terms that have subtypes and should be carefully checked to determine whether or not the current neoplasm is an additional primary or a more specific subtype.

Term	Code
Malignant lymphoma, NOS	9590/3
Malignant lymphoma, non-Hodgkin, NOS	9591/3
Hodgkin lymphoma, NOS	9650/3
Follicular lymphoma, NOS	9690/3
Leukemia, NOS	9800/3
Acute leukemia, NOS	9801/3
Myeloid leukemia, NOS	9860/3
Acute myeloid leukemia, NOS	9861/3
Chronic myeloproliferative disease, NOS	9960/3
Myeloproliferative disease, NOS	9975/3
Myelodysplastic syndrome, NOS	9989/3

Appendix F
Master Code Lists

Table F1: WHO/ICD-O-3 Master List of Histology Codes – Alphabetic List

2010 WHO Only: New histology terms and codes published in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition. These codes are not in ICD-O-3 or the ICD-O-3 errata.

ICD-O-3 Only: The histology term and code are published only in ICD-O-3

WHO and ICD-O-3: The histology term is the WHO preferred term for that neoplasm; the WHO and ICD-O-3 preferred histology terms may not be the same, but the codes for this neoplasm are published in both WHO and in ICD-O-3.

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Acute basophilic leukemia			9870/3
Acute biphenotypic leukemia		9805/3	
Acute erythroid leukemia			9840/3
Acute megakaryoblastic leukemia			9910/3
Acute monoblastic and monocytic leukemia			9891/3
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13);RBM15-MKL1	9911/3		
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22), CBFB/MYH11			9871/3
Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26;2); RPN1-EVI1	9869/3		
Acute myeloid leukemia with maturation			9874/3
Acute myeloid leukemia with minimal differentiation			9872/3
Acute myeloid leukemia with mutated CEBPA			9861/3
Acute myeloid leukemia with mutated NPM1			9861/3
Acute myeloid leukemia with myelodysplasia-related changes			9895/3
Acute myeloid leukemia with t(6;9)(p23;q34); DEK-NUP214	9865/3		
Acute myeloid leukemia with t(8;21)(q22;q22); RUNX1-RUNX1T1			9896/3
Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL			9897/3
Acute myeloid leukemia without maturation			9873/3
Acute myeloid leukemia, NOS			9861/3
Acute myelomonocytic leukemia			9867/3
Acute panmyelosis with myelofibrosis			9931/3
Acute promyelocytic leukemia (AML with t(15;17)(q22;q12), PML/RARA			9866/3
Acute undifferentiated leukemia			9801/3
Adult T-cell leukemia/lymphoma			9827/3
Aggressive NK-cell leukemia			9948/3
ALK positive large B-cell lymphoma	9737/3		
Alpha heavy chain disease			9762/3
Anaplastic large cell lymphoma, ALK negative			9702/3
Anaplastic large cell lymphoma, ALK positive			9714/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Angioimmunoblastic T-cell lymphoma			9705/3
Atypical chronic myeloid leukemia, BCR-ABL1 negative			9876/3
B lymphoblastic leukemia/lymphoma with hyperdiploid	9815/3		
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3		
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3);E2A-PBX1 (TCF3-PBX1)	9818/3		
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22);TEL-AML1 (ETV6-RUNX1)	9814/3		
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32);IL3-IGH	9817/3		
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2);BCR-ABL1	9812/3		
B lymphoblastic leukemia/lymphoma with t(v;11q23);MLL rearranged	9813/3		
B lymphoblastic leukemia/lymphoma, NOS	9811/3		
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma			9596/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma			9680/3
B-cell prolymphocytic leukemia			9833/3
Blastic plasmacytoid dendritic cell neoplasm			9727/3
Burkitt cell leukemia			9826/3
Burkitt lymphoma			9687/3
Chronic eosinophilic leukemia, NOS			9964/3
Chronic lymphocytic leukemia/small lymphocytic lymphoma			9823/3
Chronic lymphoproliferative disorder of NK-cells			9831/3
Chronic myelogenous leukemia, BCR-ABL1 positive			9875/3
Chronic myeloid leukemia, NOS			9863/3
Chronic myelomonocytic leukemia			9945/3
Chronic myeloproliferative disease, NOS			9960/3
Chronic neutrophilic leukemia			9963/3
Classical Hodgkin lymphoma			9650/3
Diffuse large B-cell lymphoma (DLBCL), NOS			9680/3
DLBCL associated with chronic inflammation			9680/3
EBV Positive DLBCL of the elderly			9680/3
Enteropathy-associated T-cell lymphoma			9717/3
Essential thrombocythemia			9962/3
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)			9699/3
Extranodal NK/T cell lymphoma, nasal type			9719/3
Extraosseous plasmacytoma			9734/3
Fibroblastic reticular cell tumor	9759/3		
Follicular dendritic cell sarcoma			9758/3
Follicular lymphoma			9690/3
Follicular lymphoma, grade 1			9695/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Follicular lymphoma, grade 2			9691/3
Follicular lymphoma, grade 3A			9698/3
Follicular lymphoma, grade 3B			9698/3
Gamma heavy chain disease			9762/3
Hairy cell leukemia			9940/3
Hairy cell leukemia-variant			9591/3
Heavy chain disease			9762/3
Hepatosplenic T-cell lymphoma			9716/3
Histiocytic sarcoma			9755/3
Hodgkin granuloma [obs]		9661/3	
Hodgkin lymphoma, lymphocyte depletion, diffuse fibrosis		9654/3	
Hodgkin lymphoma, lymphocyte depletion, reticular		9655/3	
Hodgkin lymphoma, nodular sclerosis, cellular phase		9664/3	
Hodgkin lymphoma, nodular sclerosis, grade 1		9665/3	
Hodgkin lymphoma, nodular sclerosis, grade 2		9667/3	
Hodgkin sarcoma [obs]		9662/3	
Hydroa vacciniforme-like lymphoma	9725/3		
Immunoproliferative disease, NOS		9760/3	
Immunoproliferative small intestinal disease		9764/3	
Indeterminate dendritic cell tumor			9757/3
Interdigitating dendritic cell sarcoma			9757/3
Intravascular large B-cell lymphoma	9712/3		
Juvenile myelomonocytic leukemia			9946/3
Langerhans cell histiocytos			9751/3
Langerhans cell sarcoma			9756/3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3		
Leukemia, NOS		9800/3	
Lymphocyte-depleted classical Hodgkin lymphoma			9653/3
Lymphocyte-rich classical Hodgkin lymphoma			9651/3
Lymphoid leukemia, NOS		9820/3	
Lymphoplasmacytic lymphoma			9671/3
Malignant histiocytosis		9750/3	
Malignant lymphoma, large B-cell, diffuse, immunoblastic, NOS		9684/3	
Malignant lymphoma, mixed small and large cell, diffuse [obs]		9675/3	
Malignant lymphoma, NOS		9590/3	
Malignant lymphoma, small B lymphocytic, NOS		9670/3	
Mantle cell lymphoma			9673/3
Mast cell leukemia			9742/3
Mast cell sarcoma			9740/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Mixed cellularity classical Hodgkin lymphoma			9652/3
Mixed phenotype acute leukemia with t(9;22(q34;q11.2);BCR-ABL1	9806/3		
Mixed phenotype acute leukemia with t(v;11q23);MLL, rearranged	9807/3		
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3		
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3		
Mu heavy chain disease			9762/3
Mycosis fungoides			9700/3
Myelodyasplastic syndrome associated with isolated del(5q)			9986/3
Myelodysplasic syndrome, unclassifiable			9989/3
Myelodysplastic/myeloproliferative neoplasm, unclassifiable			9975/3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3		
Myeloid and lymphoid neoplasm with PDGFRA rearrangement	9965/3		
Myeloid leukemia associated with Down syndrome	9898/3		
Myeloid leukemia, NOS			9860/3
Myeloid neoplasm with PDGFRB arrangement	9966/3		
Myeloid sarcoma			9930/3
Myeloproliferative neoplasm, unclassifiable			9975/3
Nodal marginal zone lymphoma			9699/3
Nodular lymphocyte predominant Hodgkin lymphoma			9659/3
Nodular sclerosis classical Hodgkin lymphoma			9663/3
Pediatric follicular lymphoma			9690/3
Pediatric nodal marginal zone lymphoma			9699/3
Peripheral T-cell lymphoma, NOS			9702/3
Plasma cell leukemia			9733/3
Plasma cell myeloma			9732/3
Plasmablastic lymphoma	9735/3		
Polycythemia vera			9950/3
Polymorphic Post Transplant Lymphoproliferative Disorder (PTLD)	9971/3		
Precursor B-cell lymphoblastic leukemia/lymphoma			9836/3
Precursor B-cell lymphoblastic lymphoma		9728/3	
Precursor cell lymphoblastic leukemia, NOS			9835/3
Precursor T-cell lymphoblastic lymphoma			9729/3
Primary cutaneous anaplastic large cell lymphoma			9718/3
Primary cutaneous CD4 positive small/medium cell T-cell lymphoma			9709/3
Primary cutaneous CD8 positive aggressive epidermotropic cytotoxic T-cell lymphoma			9709/3
Primary cutaneous DLBCL, leg type			9680/3
Primary cutaneous follicle centre lymphoma	9597/3		
Primary cutaneous gamma-delta T-cell lymphoma	9726/3		
Primary DLBCL of the CNS,			9680/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Primary effusion lymphoma			9678/3
Primary mediastinal (thymic) large B-cell lymphoma			9679/3
Primary myelofibrosis			9961/3
Prolymphocytic leukemia, NOS			9832/3
Refractory anemia			9980/3
Refractory anemia with excess blasts			9983/3
Refractory anemia with excess blasts in transformation [obs]			9984/3
Refractory anemia with ring sideroblasts			9982/3
Refractory anemia with ring sideroblasts associated with marked thrombocytosis			9982/3
Refractory cytopenia of childhood			9985/3
Refractory cytopenia with multilineage dysplasia			9985/3
Refractory neutropenia	9991/3		
Refractory thrombocytopenia	9992/3		
Sezary syndrome			9701/3
Solitary plasmacytoma of bone			9731/3
Splenic B-cell lymphoma/leukemia, unclassifiable			9591/3
Splenic B-cell marginal zone lymphoma			9689/3
Splenic diffuse red pulp small B-cell lymphoma			9591/3
Subcutaneous panniculitis-like T-cell lymphoma			9708/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3		
Systemic mastocytosis			9741/3
T lymphoblastic leukemia/lymphoma			9837/3
T-cell large granular lymphocytic leukemia			9831/3
T-cell prolymphocytic leukemia			9834/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3		
Therapy-related myelodysplastic syndrome, NOS			9987/3
Therapy-related myeloid neoplasm			9920/3
Waldenstrom macroglobulinemia			9761/3

Table F2: WHO/ICD-O-3 Master List of Histology Codes – Numeric List

2010 WHO Only: New histology terms and codes published in the *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition. These codes are not in ICD-O-3 or the ICD-O-3 errata.

ICD-O-3 Only: The histology term and code are published only in ICD-O-3

WHO and ICD-O-3: The histology term is the WHO preferred term for that neoplasm; the WHO and ICD-O-3 preferred histology terms may not be the same, but the codes for this neoplasm are published in both WHO and in ICD-O-3.

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Malignant lymphoma, NOS		9590/3	
Hairy cell leukemia-variant			9591/3
Splenic B-cell lymphoma/leukemia, unclassifiable			9591/3
Splenic diffuse red pulp small B-cell lymphoma			9591/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma			9596/3
Primary cutaneous follicle centre lymphoma	9597/3		
Classical Hodgkin lymphoma			9650/3
Lymphocyte-rich classical Hodgkin lymphoma			9651/3
Mixed cellularity classical Hodgkin lymphoma			9652/3
Lymphocyte-depleted classical Hodgkin lymphoma			9653/3
Hodgkin lymphoma, lymphocyte depletion, diffuse fibrosis		9654/3	
Hodgkin lymphoma, lymphocyte depletion, reticular		9655/3	
Nodular lymphocyte predominant Hodgkin lymphoma			9659/3
Hodgkin granuloma [obs]		9661/3	
Hodgkin sarcoma [obs]		9662/3	
Nodular sclerosis classical Hodgkin lymphoma			9663/3
Hodgkin lymphoma, nodular sclerosis, cellular phase		9664/3	
Hodgkin lymphoma, nodular sclerosis, grade 1		9665/3	
Hodgkin lymphoma, nodular sclerosis, grade 2		9667/3	
Malignant lymphoma, small B lymphocytic, NOS		9670/3	
Lymphoplasmacytic lymphoma			9671/3
Mantle cell lymphoma			9673/3
Malignant lymphoma, mixed small and large cell, diffuse [obs]		9675/3	
Primary effusion lymphoma			9678/3
Primary mediastinal (thymic) large B-cell lymphoma			9679/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma			9680/3
Diffuse large B-cell lymphoma (DLBCL), NOS			9680/3
DLBCL associated with chronic inflammation			9680/3
EBV Positive DLBCL of the elderly			9680/3
Primary cutaneous DLBCL, leg type			9680/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Primary DLBCL of the CNS,			9680/3
Malignant lymphoma, large B-cell, diffuse, immunoblastic, NOS		9684/3	
Burkitt lymphoma			9687/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3		
Splenic B-cell marginal zone lymphoma			9689/3
Follicular lymphoma			9690/3
Pediatric follicular lymphoma			9690/3
Follicular lymphoma, grade 2			9691/3
Follicular lymphoma, grade 1			9695/3
Follicular lymphoma, grade 3A			9698/3
Follicular lymphoma, grade 3B			9698/3
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)			9699/3
Nodal marginal zone lymphoma			9699/3
Pediatric nodal marginal zone lymphoma			9699/3
Mycosis fungoides			9700/3
Sezary syndrome			9701/3
Anaplastic large cell lymphoma, ALK negative			9702/3
Peripheral T-cell lymphoma, NOS			9702/3
Angioimmunoblastic T-cell lymphoma			9705/3
Subcutaneous panniculitis-like T-cell lymphoma			9708/3
Primary cutaneous CD4 positive small/medium cell T-cell lymphoma			9709/3
Primary cutaneous CD8 positive aggressive epidermotropic cytotoxic T-cell lymphoma			9709/3
Intravascular large B-cell lymphoma	9712/3		
Anaplastic large cell lymphoma, ALK positive			9714/3
Hepatosplenic T-cell lymphoma			9716/3
Enteropathy-associated T-cell lymphoma			9717/3
Primary cutaneous anaplastic large cell lymphoma			9718/3
Extranodal NK/T cell lymphoma, nasal type			9719/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3		
Hydroa vacciniforme-like lymphoma	9725/3		
Primary cutaneous gamma-delta T-cell lymphoma	9726/3		
Blastic plasmacytoid dendritic cell neoplasm			9727/3
Precursor B-cell lymphoblastic lymphoma		9728/3	
Precursor T-cell lymphoblastic lymphoma			9729/3
Solitary plasmacytoma of bone			9731/3
Plasma cell myeloma			9732/3
Plasma cell leukemia			9733/3
Extraosseous plasmacytoma			9734/3
Plasmablastic lymphoma	9735/3		

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
ALK positive large B-cell lymphoma	9737/3		
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3		
Mast cell sarcoma			9740/3
Systemic mastocytosis			9741/3
Mast cell leukemia			9742/3
Malignant histiocytosis		9750/3	
Langerhans cell histiocytos			9751/3
Histiocytic sarcoma			9755/3
Langerhans cell sarcoma			9756/3
Indeterminate dendritic cell tumor			9757/3
Interdigitating dendritic cell sarcoma			9757/3
Follicular dendritic cell sarcoma			9758/3
Fibroblastic reticular cell tumor	9759/3		
Immunoproliferative disease, NOS		9760/3	
Waldenstrom macroglobulinemia			9761/3
Alpha heavy chain disease			9762/3
Gamma heavy chain disease			9762/3
Heavy chain disease			9762/3
Mu heavy chain disease			9762/3
Immunoproliferative small intestinal disease		9764/3	
Leukemia, NOS		9800/3	
Acute undifferentiated leukemia			9801/3
Acute biphenotypic leukemia		9805/3	
Mixed phenotype acute leukemia with t(9;22(q34;q11.2);BCR-ABL1	9806/3		
Mixed phenotype acute leukemia with t(v;11q23);MLL, rearranged	9807/3		
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3		
Mixed phenotype acute leukemia, T/myeloid, NOS	9809/3		
B lymphoblastic leukemia/lymphoma, NOS	9811/3		
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2);BCR-ABL1	9812/3		
B lymphoblastic leukemia/lymphoma with t(v;11q23);MLL rearranged	9813/3		
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22);TEL-AML1 (ETV6-RUNX1)	9814/3		
B lymphoblastic leukemia/lymphoma with hyperdiploid	9815/3		
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3		
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32);IL3-IGH	9817/3		
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3);E2A-PBX1 (TCF3-PBX1)	9818/3		
Lymphoid leukemia, NOS		9820/3	
Chronic lymphocytic leukemia/small lymphocytic lymphoma			9823/3
Burkitt cell leukemia			9826/3
Adult T-cell leukemia/lymphoma			9827/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Chronic lymphoproliferative disorder of NK-cells			9831/3
T-cell large granular lymphocytic leukemia			9831/3
Prolymphocytic leukemia, NOS			9832/3
B-cell prolymphocytic leukemia			9833/3
T-cell prolymphocytic leukemia			9834/3
Precursor cell lymphoblastic leukemia, NOS			9835/3
Precursor B-cell lymphoblastic leukemia/lymphoma			9836/3
T lymphoblastic leukemia/lymphoma			9837/3
Acute erythroid leukemia			9840/3
Myeloid leukemia, NOS			9860/3
Acute myeloid leukemia with mutated CEBPA			9861/3
Acute myeloid leukemia with mutated NPM1			9861/3
Acute myeloid leukemia, NOS			9861/3
Chronic myeloid leukemia, NOS			9863/3
Acute myeloid leukemia with t(6;9)(p23;q34); DEK-NUP214	9865/3		
Acute promyelocytic leukemia (AML with t(15;17)(q22;q12), PML/RARA			9866/3
Acute myelomonocytic leukemia			9867/3
Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26;2); RPN1-EVI1	9869/3		
Acute basophilic leukemia			9870/3
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22), CBFB/MYH11			9871/3
Acute myeloid leukemia with minimal differentiation			9872/3
Acute myeloid leukemia without maturation			9873/3
Acute myeloid leukemia with maturation			9874/3
Chronic myelogenous leukemia, BCR-ABL1 positive			9875/3
Atypical chronic myeloid leukemia, BCR-ABL1 negative			9876/3
Acute monoblastic and monocytic leukemia			9891/3
Acute myeloid leukemia with myelodysplasia-related changes			9895/3
Acute myeloid leukemia with t(8;21)(q22;q22); RUNX1-RUNX1T1			9896/3
Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL			9897/3
Myeloid leukemia associated with Down syndrome	9898/3		
Acute megakaryoblastic leukemia			9910/3
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13);RBM15-MKL1	9911/3		
Therapy-related myeloid neoplasm			9920/3
Myeloid sarcoma			9930/3
Acute panmyelosis with myelofibrosis			9931/3
Hairy cell leukemia			9940/3
Chronic myelomonocytic leukemia			9945/3
Juvenile myelomonocytic leukemia			9946/3
Aggressive NK-cell leukemia			9948/3

Histologic Term	2010 WHO Only	ICD-O-3 Only	WHO and ICD-O-3
Polycythemia vera			9950/3
Chronic myeloproliferative disease, NOS			9960/3
Primary myelofibrosis			9961/3
Essential thrombocythemia			9962/3
Chronic neutrophilic leukemia			9963/3
Chronic eosinophilic leukemia, NOS			9964/3
Myeloid and lymphoid neoplasm with PDGFRA rearrangement	9965/3		
Myeloid neoplasm with PDGFRB arrangement	9966/3		
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3		
Polymorphic Post Transplant Lymphoproliferative Disorder (PTLD)	9971/3		
Myelodysplastic/myeloproliferative neoplasm, unclassifiable			9975/3
Myeloproliferative neoplasm, unclassifiable			9975/3
Refractory anemia			9980/3
Refractory anemia with ring sideroblasts			9982/3
Refractory anemia with ring sideroblasts associated with marked thrombocytosis			9982/3
Refractory anemia with excess blasts			9983/3
Refractory anemia with excess blasts in transformation [obs]			9984/3
Refractory cytopenia of childhood			9985/3
Refractory cytopenia with multilineage dysplasia			9985/3
Myelodyasplastic syndrome associated with isolated del(5q)			9986/3
Therapy-related myelodysplastic syndrome, NOS			9987/3
Myelodysplasic syndrome, unclassifiable			9989/3
Refractory neutropenia	9991/3		
Refractory thrombocytopenia	9992/3		