Hematopoietic and Lymphoid Neoplasm Coding Manual

Effective with Cases Diagnosed 1/1/2010 and Forward

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Dedication

The Hematopoietic and Lymphoid Neoplasm Coding Manual and the companion Hematopoietic and Lymphoid Neoplasm Database (Heme DB) are dedicated to the hard-working cancer registrars across the world who meticulously identify, abstract, and code cancer data. Cancer registrars are the foundation for statewide, provincial, territorial, national, and international cancer surveillance programs which support cancer prevention and cancer control efforts worldwide.

Education

SEER*Educate provides training on how to use the Heme Manual and DB. Step-by-step instructions are provided for each case scenario to learn how to use the application and manual to arrive at the answer provided.

Use this manual and the corresponding database to abstract and code cases diagnosed **January 1, 2010** and forward. Some information for cases diagnosed prior to 2010 is also provided to assist registrars in coding those cases and in making multiple primary decisions.

Hematopoietic Comparison Documents
Starting January 1, 2014, only one database for diagnosis years 2010 and forward is available and is referred to as the "Hematopoietic and Lymphoid Neoplasm Database." The consolidated manual and database include changes from 2010, 2012 and 2014. Earlier versions (2010 and 2012) are no longer available.

Comparison documents have been developed to identify the differences between the 2010, 2012 and 2014 changes of the Hematopoietic manual and database. These documents are meant to be a guide for registrars on how their data may have changed over time. Included with the rules is a column that comments on possible changes to incidence. Most of the changes will not affect incidence. There is an explanation of how and what to review where incidence may be affected. The documents can be found at: http://www.seer.cancer.gov/tools/heme/comparison.html

Versions
2010 Hematopoietic Coding Manual and Database (Effective dates 1/1/2010-12/31/2011)
- Release date: March 2010, Version 1.4 (initial release)
- Release date: April 2010, Version 1.5
- Release date: June 2010, Version 1.6

2012 Hematopoietic Coding Manual and Database (Effective dates 1/1/2012-12/31/2013)
- Release date: May 4, 2012, Version 2.1
- Release date: May 23, 2012, Version 2.1
- Release date: February 25, 2013, Version 2.2

Hematopoietic and Lymphoid Neoplasm Coding Manual (Effective 1/1/2010)
- Release date: January 2014

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- Release date: January 2015

2015 Revisions
The 2015 revisions include
- Rule clarifications and revisions
- Correction of typos (in both manual and database)
- Expanded treatment information
- Expanded ambiguous terminology information
- Non-reportable terms removed from Hematopoietic Manual Appendix F and added to the database
- Glossary removed from the Manual and entered into the new Glossary database
- A complete list of changes are available on the SEER website

Many of the revisions are based on questions submitted to Ask a SEER Registrar. Selected questions and answers from Ask a SEER Registrar are posted in SINQ, which is updated on a regular basis.

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Introduction and 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 Heme DB reference to assist registrars in determining case reportability, the number of primaries, as well as instructions for coding primary site, histology, grade, diagnostic confirmation and other therapy for a hematopoietic and/or lymphoid neoplasm. The rules, guidelines, and the Hematopoietic DB follow the *World Health Organization (WHO) Classification of Tumours of 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 World Health Organization (WHO), but the content of the books are very different. Each has a prominent place in the oncology 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 and historic cases. When ICD-O assigns a code to a specific histology, the original code is rarely 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. The ICD-O 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, that process 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 numeric position in ICD-O. When the ICD-O editors assign new codes for a neoplasm, the new code may not be placed in the desired category because there may not be room within that category to add a new code. 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 Hematopathology 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 neoplasms. This reference was the primary source of information used to develop the *2010 Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual*, the *2012 Hematopoietic Coding Manual*, and the accompanying Hematopoietic DB because the WHO Blue Book is periodically 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 current pathology reports and 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 in categories based on information about the phenotype or behavior of the neoplasm.

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/)

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

The classification of the leukemias and lymphomas can be confusing because of the variety of cell types involved, the site of origin of the neoplastic process (bone marrow, lymph node, GI tract, etc.), and the relative frequency or infrequency of tumor cells circulating in the peripheral blood.

Leukemia and lymphoma are terms that reflect the primary behavior and often the primary site of a neoplasm. Leukemias have cells circulating in the peripheral blood, which can originate in lymph nodes or the marrow. Lymphomas generally form solid masses in lymph nodes or organs containing lymphoid tissue; they may occasionally have circulating tumor cells as well.

Leukemias and lymphomas may also be defined as being chronic or acute. Chronic neoplasms are of longer duration and are slowly progressive while acute neoplasms are of shorter duration and rapidly progressing.

Some examples of chronic and acute:

Cell Type	Chronic	Acute
Lymphocyte	Chronic lymphocytic leukemia/small lymphocytic leukemia (CLL/SLL); follicular lymphoma; diffuse large B-cell lymphoma, plasma cell myeloma	Lymphoblastic leukemia (T or B cell)
Granulocyte	Chronic myeloid leukemia	Acute myeloid leukemia
Erythrocyte	Polycythemia vera	Acute erythroid leukemia
Megakaryocytic	Essential thrombocythemia	Acute megakaryocytic leukemia

Leukemias and Lymphomas

Leukemia vs. Lymphoma

One of the differences 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 diagnosis is usually leukemia. Although rare, a lymphoma may present only in the bone marrow. (See PH rules, Modules 6 and 7, 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. The spleen filters and stores blood cells. The spleen involvement is usually secondary, much like metastases in solid tumors. The rare histologies that are primary in the spleen are identified in the Heme DB. The Primary Site will be listed as C422.

Diagnostic Process for Leukemia

For most patients, the first suspicion or presentation of a hematopoietic neoplasm will be symptoms such as unexplained weight loss, weakness, chronic fatigue, easy bruising, etc. When the physician suspects leukemia, he/she usually orders a complete blood count (CBC) and/or a peripheral blood smear. The CBC will identify abnormalities of the platelets, hemoglobin, white blood cells or red blood cells. When an abnormality is identified in the blood cell analysis, a bone marrow (BM) biopsy is usually the next procedure. The CBC or bone marrow alone seldom provide a definitive diagnosis; however, the results usually provide one or more provisional diagnoses such as: myeloproliferative neoplasms, myeloid neoplasms, myelodysplastic/myeloproliferative neoplasms, myelodysplastic syndromes, or leukemia. These non-specific diagnoses are differential or provisional. More testing is needed to identify the specific hematopoietic or lymphoid neoplasm. Many of the neoplasms in the 2008 WHO Classification require immunophenotyping or genetic information to identify the specific histology. The Heme DB contains information on the types of diagnostic tests that are used to identify the specific histology for the hematopoietic or lymphoid neoplasm being abstracted. See the "Definitive Diagnostic Method" section in the Heme DB.

Lymphoma

Biopsies

The most accessible involved lymph node or site is usually biopsied when lymphoma is suspected. For example, if a CT or PET scan identified enlarged cervical and mediastinal lymph nodes, the physician would biopsy the cervical lymph nodes because that would be the least invasive procedure; i.e. the cervical nodes are more accessible than the mediastinal nodes. Do **not** assume that the more accessible site chosen for biopsy is the primary site. Follow the primary site rules and instructions when coding Primary Site.

Hodgkin Lymphoma

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 neoplasm from one lymph node chain to another. The neoplasm may progress to involve the spleen, liver, and/or bone marrow.

Non-Hodgkin Lymphoma

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

Note: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/3-9653/3, 9655/3, 9659/3, 9663/3

Myeloid neoplasms

WHO lists the following major subgroups of Myeloid neoplasms:

- Acute myeloid leukemia (AML)
- Myelodysplastic syndromes (MDS)
- Myelodysplastic/myeloproliferative neoplasms (MDS/MPN)
- Myeloid and lymphoid neoplasms with eosinophilia and abnormalities of PDGFRA, PDGFRB, or FGFR1
- Myeloproliferative neoplasms (MPN)

MDS are among the most challenging of the myeloid neoplasms for diagnosis and classification. MDS include the following:.

- Childhood myelodysplastic syndrome
- Myelodysplastic syndrome with isolated del(5q)
- Myelodysplastic syndrome, unclassifiable (MDS, U)
- Refractory anemia with excess blasts (RAEB)
- Refractory anemia with ring sideroblasts (RARS)
- Refractory cytopenia with multilineage dysplasia (RCMD)
- Refractory cytopenia with unilineage dysplasia (RCUD)
    - Refractory anemia (RA)
    - Refractory neutropenia (RN)
    - Refractory thrombocytopenia (RT)

MDS/MPN present with findings supporting a diagnosis of MDS and other findings supporting a diagnosis of MPN. MDS/MPN include the following:

- Atypical chronic myeloid leukemia BCR-ABL1 negative (aCML)
- Chronic myelomonocytic leukemia (CMML)
- Juvenile myelomonocytic leukemia (JMML)
- Myelodysplastic/myeloproliferative neoplasm, unclassifiable (MDS/MPN, U)

MPN are stem cell disorders characterized by proliferation of one or more myeloid linage. MPNs include the following:

- Chronic eosinophilic leukemia, NOS (CEL, NOS)
- Chronic myelogenous leukemia, BCR-ABL positive (CML)
- Chronic neutrophilic leukemia (CNL)
- Essential thrombocythemia (ET)
- Mastocytosis
- Myeloproliferative neoplasm, unclassifiable (MPN, U)
- Polycythemia vera (PV)
- Primary myelofibrosis (PMF)

The Hematopoietic Database (Heme DB)

The Heme DB is available online through the SEER website. Access the database at http://seer.cancer.gov/seertools/hemelymph/. An internet connection is required. Please note that the online version cannot be downloaded onto a PC or laptop.

A stand-alone version of the Heme DB that can be downloaded onto PCs and laptops is available for those who cannot access the online version. At some point in the future, only the online version will be available in order to eliminate costs associated with maintaining two formats. Please note that the stand-alone version is updated less often than the online version.

The Heme DB enables registrars to identify and understand hematopoietic and lymphoid neoplasms as well as to correctly and consistently abstract and code cases. Users are able to query any final, differential, or provisional diagnosis in the Heme DB. The diagnostic or confirmatory tests are listed under "Definitive Diagnostic Methods" for each neoplasm. The information needed to search the medical record for specific diagnostic test results is provided. Some healthcare institutions may "file" confirmatory test results, such as immunophenotyping or genetic testing, in a location other than that used for standard laboratory tests in the medical record. We recommend that the registrar ask the laboratory for examples of test results, such as immunophenotyping or genetic testing, to become familiar with the test names and format of the test results as well as other information that may be included with the lab analysis. 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).

Crosswalk

For the histology codes introduced in 2010, information is now available in the Heme database in the Abstractor Notes section stating which code(s) would have been used prior to 2010 to code the diagnosis.

Coding the Data Item Diagnostic Confirmation (NAACCR Item #490)

Codes for Hematopoietic and Lymphoid Neoplasms (9590/3-9992/3)

Microscopically Confirmed

Code	Description
1	Positive histology
2	Positive cytology
3	Positive histology PLUS: • Positive immunophenotyping AND/OR • Positive genetic studies (***Effective for cases diagnosed 1/1/2010 and later)***

Not Microscopically Confirmed

Code	Description
5	Positive laboratory test/marker study
6	Direct visualization without microscopic confirmation
7	Radiology and other imaging techniques without microscopic confirmation
8	Clinical diagnosis only (other than 5, 6 or 7)

Confirmation Unknown

Code	Description
9	Unknown whether or not microscopically confirmed; death certificate only

Coding Instructions continued on next page

Diagnostic Confirmation Coding Instructions for Hematopoietic and Lymphoid Neoplasms (9590/3-9992/3)

Note 1: There is no priority order or hierarchy for coding the Diagnostic Confirmation for hematopoietic or lymphoid neoplasms. Most commonly the bone marrow provides several provisional diagnoses and the specific histologic type is determined through immunophenotyping or genetic testing.

Note 2: Use code 1 when **ONLY** the tissue, bone marrow, or blood was used to diagnose the specific histology. Do **not** use code 1 if the provisional diagnosis was based on tissue, bone marrow, or blood **and** the immunophenotyping or genetic testing on that same tissue, bone marrow, or blood identified the specific disease (see Code 3).

Note 3: If a neoplasm is originally confirmed by histology (code 1), and later has immunophenotyping, genetic testing or JAK2 which confirms a more specific neoplasm and there is no evidence of transformation, change the histology code to the more specific neoplasm and change the diagnostic confirmation to code 3.
- Do **not** use diagnostic confirmation code 3 for cases diagnosed prior to 1/1/2010.

Code 1: Positive histology

Code 1 includes a provisional diagnosis and/or several provisional (differential) diagnoses which may or may not be preceded by approved ambiguous terminology.

Assign code 1 for

1. Tissue from lymph node(s), organ(s) or other tissue specimens from biopsy, frozen section, surgery, or autopsy
2. Bone marrow specimens (aspiration and biopsy)
3. Peripheral blood smear
    a. Can be used as a histological diagnoses for any of the hematopoietic histologies (9590/3-9992/3)
4. L**eukemia** only (9800/3-9948/3), positive histology also includes
    a. Complete blood count (CBC)
    b. White blood count (WBC)
5. Neoplasm microscopically confirmed **AND**
    a. Immunophenotyping, genetic testing, or JAK2 **not** done **OR**
    b. Immunophenotyping, genetic testing, or JAK2 done but **negative** (non-diagnostic) for the neoplasm being abstracted

    Example: Acute myelomonocytic leukemia (9867/3) CD10+. CD10+ is not listed under Immunophenotyping for this histology, so diagnostic confirmation should be 1.
6. Use for historical cases not already in the database if information states that there was histologic confirmation

    Example: Patient diagnosed in 2012 with Stage III mantle cell lymphoma, diagnosed by LN biopsy. Mantle cell lymphoma not in the database. Now presents with DLBCL in 2015.

Code 2: Positive cytology

Code 2 is rarely used for Hematopoietic and Lymphoid neoplasms.

Assign code 2 for

1. Examination of fluid such as spinal fluid, peritoneal fluid, or pleural fluid
2. Paraffin block specimens from concentrated spinal fluid, peritoneal fluid, or pleural fluid
3. A specimen that fails to provide enough tissue to do a histologic examination - in this case, the report will be a cytology report rather than a pathology report

Code 3: Positive histology PLUS positive immunophenotyping or genetic testing

Code 3 can be used for cases diagnosed **2010+** with histologic confirmation (see code 1) **AND** immunophenotyping, genetic testing, or JAK2 confirmation

Assign code 3 for

1. Cases positive for neoplasm being abstracted (including acceptable ambiguous terminology and provisional diagnosis) **AND** Immunophenotyping, genetic testing, or JAK2 is listed in the Definitive Diagnosis in the Heme DB **AND**
    a. Confirms the neoplasm **OR**
    b. Identifies a more specific histology (not preceded by ambiguous terminology)
    Note 1: Do not use code 3 for positive immunophenotyping or genetic testing identifying a more specific histology when preceded by ambiguous terminology.
    Note 2: Do not use code 3 for positive immunophenotyping or genetic testing identifying a more specific histology when the test result is preceded by "patchy weak staining."

Example 1 (Identifying a more specific histology): Bone marrow biopsy positive for acute myeloid leukemia (9861/3). Genetic testing positive for AML with inv (16) (p13.1q22) (9871/3). Code Diagnostic Confirmation code 3, positive histology and positive genetic testing.

Example 2 (Identifying a more specific histology): Peripheral blood smear with lymphoblastic lymphoma (9671/3). Bone marrow biopsy with immunophenotyping showing CD5 negative and IgM positive, diagnosis Waldenstrom Macroglobulinemia (9761/3). Code Diagnostic Confirmation code 3, positive histology and positive immunophenotyping testing which identified a more specific histology.

Example 3 (Confirming the histologic diagnosis): Bone marrow biopsy diagnosis is plasma cell dyscrasia (9765/1). Peripheral blood smear is compatible with plasma cell leukemia. FISH and chromosome analysis revealed plasma cell myeloma. Both plasma cell leukemia and plasma cell myeloma are coded to the same ICD-O-3 code, 9732/3, so there is only one disease process. The peripheral blood smear is histologic diagnosis for the plasma cell leukemia and FISH confirmed the diagnosis of multiple myeloma/plasma cell myeloma. Code Diagnostic Confirmation 3, positive histology and positive genetic testing.

Example 4 (Histologic confirmation plus genetic and immunophenotyping confirmation): Patient diagnosed with CLL by CBC and flow cytometry that was positive for both the genetic and CD antigens (immunophenotyping) for CLL. A bone marrow biopsy not performed. Since this is leukemia, the CBC is histologic confirmation, so this patient had histologic confirmation, genetic, and immunophenotyping positive for CLL. Code Diagnostic Confirmation 3, positive histology and positive genetic testing/immunophenotyping.

Example 5 (Ambiguous terminology used with immunophenotyping): Bone marrow biopsy shows B lymphoblastic leukemia. Abnormal FISH results most likely represent a hyperdiploid clone. Code the histology to 9811 (B-ALL, NOS) and assign a diagnostic confirmation code of 1. Neither Diagnostic confirmation 3 nor the more specific hyperdiploidy histology is coded because the associated FISH result is preceded by ambiguous terminology.

Code 4: Positive microscopic confirmation, method not specified

Code 4 is rarely used for Hematopoietic and Lymphoid neoplasms.

1. Assign code 4 when there is information that the diagnosis of cancer was microscopically confirmed, but the type of confirmation is unknown

Code 5: Positive laboratory test/marker study

Code 5 is rarely used for Hematopoietic and Lymphoid neoplasms. If there was no provisional diagnosis or clinical suspicion of cancer, immunophenotyping or genetic testing would not be done.

Example: CT scan consistent with multiple myeloma (9732/3). Twenty-four hour urine protein elevated with the presence of Bence-Jones kappa. Code 5 for diagnosis based on the positive Bence-Jones, which is listed as one of the diagnostic confirmation methods in the Heme DB and is also a lab test.

Code 6: Direct visualization without microscopic confirmation
Code 6 is rarely used for Hematopoietic and Lymphoid neoplasms.
Assign code 6 when

1. The operative report states the patient had lymphoma, but no biopsy or cytology was done
2. The diagnosis is determined by gross autopsy findings (no tissue or cytologic confirmation)

Code 7: Radiology and other imaging techniques without microscopic confirmation
Code 7 is rarely used for Hematopoietic and Lymphoid neoplasms.
Assign code 7 when

1. The diagnosis is confirmed by radiology or other imaging techniques only

***Example**:* Terminally ill patient who has a CT scan with the impression: suspicious for lymphoma. The patient refused further workup.

Code 8: Clinical diagnosis only (other than 5, 6, or 7)
Assign code 8 when

1. While clinical diagnosis is seldom used for solid tumors, it is a valid diagnostic method for certain hematopoietic neoplasms.
2. The Heme DB will list Clinical Diagnosis as the definitive diagnostic method for certain hematopoietic neoplasms. For these neoplasms, biopsy, immunophenotyping, and genetic testing do not confirm the neoplasm.
3. The diagnosis is determined based on the physician's clinical expertise, combined with the information from the biopsy, equivocal or negative tests, and the clinical symptoms. This is called a "diagnosis of exclusion" because the physician's judgment and the work-up literally exclude all other possible diagnoses, leaving one diagnosis. Ambiguous terminology may precede the diagnosis.

Example: Bone marrow biopsy shows anemia NOS; physician notes states the patient's overall clinical presentation of hypercalcemia, fever, and anemia is consistent with Myelodysplastic Syndrome, NOS (9989/3). Code Diagnostic Confirmation 8, clinical diagnosis only.

Code 9: Unknown whether or not microscopically confirmed; death certificate only
Assign code 9 when

1. It is unknown if the diagnosis was confirmed microscopically
2. For death-certificate-only (DCO) cases
3. For historical cases not already in the database when there is no information available
   Example: "History of follicular lymphoma in 2010, now presents with DLBCL." Follicular lymphoma not in the database. Assign diagnostic confirmation of 9 for the follicular lymphoma.

Transformations: Chronic Neoplasms and Acute Neoplasms

Transformations to

The "Transformations" field has been relabeled "Transformations to." If a chronic neoplasm can transform to an acute/more severe neoplasm, the Heme DB will show the acute neoplasm in the "Transformations to" section. For example, if you search the Heme DB for chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL) (9823/3), the "Transformations to" section shows that CLL/SLL transforms to diffuse large B-cell lymphoma (9680/3). That means CLL/SLL is a chronic neoplasm and diffuse large B-cell lymphoma is an acute neoplasm.

Transformations from

A new field has been added to the database: "Transformations from." This field was added to help registrars determine which histologies are chronic and which are acute. Acute neoplasms may have other histologies listed in the "Transformations from" field. Histologies listed in the "Transformations from" field are chronic. For example, in the Heme DB under plasma cell myeloma (9732/3), the "Transformations from" field lists solitary plasmacytoma of bone (9731/3) and extraosseous plasmacytoma (9734/3). That means that plasma cell myeloma (9732/3) is an acute neoplasm which could have transformed from the two listed plasmacytomas (9731/3 and 9734/3) which are chronic neoplasms.

See Rules M8-M11 for determination of single or multiple primaries involving cases noting both chronic and acute diagnoses.
The most common form of transformation is when a neoplasm progresses from chronic to acute; however, neoplasms may be diagnosed in an acute phase and transform to a less aggressive chronic phase after treatment. In these cases, it is important to determine if the patient received treatment for the acute neoplasm. If the patient was treated, abstract the chronic neoplasm as a second primary (see Rule M13). If the patient was not treated for the acute neoplasm, code only the acute neoplasm (see Rule M12). Follow back is definitely recommended to determine whether there was any further diagnostic workup that proved the acute diagnosis was incorrect or documentation that the acute diagnosis was provisional.

Obsolete Hematopoietic Histology Codes

NOTE: OBS codes will no longer be used starting in January 2015 for cases diagnosed 2010 and forward. A conversion program has been developed which will replace all OBS codes with diagnosis date of 1/1/2010 or later with the current code. (See Appendix E for a listing of the codes). For complete details on the conversion program, please see http://seer.cancer.gov/tools/heme/conversion.html.

Defining obsolete codes in the Hematopoietic Database and Manual

Terminology for, and understanding of, hematopoietic neoplasms evolves and changes. The 2008 *WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues (2008 WHO)* is the primary resource used for the Heme DB and manual because it incorporates the most recent scientific understanding of these neoplasms and their classification. The *2008 WHO* reclassified some hematopoietic and lymphoid neoplasm terms and introduced some new ICD-O-3 histology codes. As a result, some histology codes have become obsolete. Terms associated with obsolete codes have been reclassified in the 2008 WHO either by moving them to an existing code or to one of the new codes. See Appendix E for the list of obsolete codes and the current codes to be used in place of the obsolete code. This information is also in the database.

Do not use obsolete histology codes for cancer registry data collection beginning with cases diagnosed 1/1/2010. In the Hematopoietic & Lymphoid Neoplasm DB, obsolete codes are identified by an "Obsolete" tag on the search results page and also by a note at the top of the disease entity explaining that the code is obsolete. Instructions for the current code are provided after the phrase "For current cases please see code…" Use the current code in place of the obsolete code. **Obsolete codes are listed in ICD-O-3; [obs] is found at end of code documentation in both the Numerical and Alphabetic indexes without further information. The obsolete codes listed in ICD-O-3 and the obsolete codes listed in the Hematopoietic & Lymphoid Neoplasm Coding Manual and Database are not the same; therefore it is important for registrars to use the Hematopoietic and Lymphoid Neoplasm Database and Coding Manual to determine the correct histology code**. Obsolete codes will continue to appear in cancer reporting software, legacy data groupings, and data reports.

> ***Note:*** Instructions in this manual for coding obsolete codes pertain to hematopoietic and lymphoid neoplasms (9590/3-9992/3) only. These instructions do not apply to solid tumors.

When to use an obsolete code

Obsolete codes in the database have the following instruction: "This code may be used for cases diagnosed prior to 1/1/2010." Use the current code for all cases diagnosed 1/1/2010 or later. Use obsolete codes only for cases diagnosed prior to 2010. **This is a change from previous versions of the manual** which stated that the obsolete code could be used for cases diagnosed 1/1/2010 or later for DCOs or cases with minimal information (path only cases).

Cases diagnosed prior to 2010

For cases diagnosed prior to 2010, look up the histologic term in the Heme DB and assign the obsolete code.

> ***Example 1:*** Pathology report dated 7/12/2009 states therapy related myelodysplastic syndrome. Code histology to 9987/3 since the date of diagnosis is prior to 1/1/2010. (Histology code 9987/3 became obsolete 1/1/2010).
>
> ***Example 2:*** Pathology report dated 12/15/2009 states malignant lymphoma, large B-cell, diffuse immunoblastic, NOS. Code histology to 9684/3 since the date of diagnosis is prior to 1/1/2010. (Histology code 9684/3 became obsolete as of 1/1/2010)

Steps in Priority Order for Using the Heme DB and Hematopoietic Coding Manual

1. Identify the working (preliminary) **histology code(s)**
    a. Search the Heme DB using a unique word in the diagnosis, for example "precursor" if the diagnosis is precursor acute lymphoblastic leukemia
    b. Or you can search on the complete name (diagnosis). For example, "acute myelomonocytic leukemia". The number of matched terms that are displayed will be much smaller than just searching on "leukemia".
        i. The search engine will display every entry with **all** of the words "acute" "myelomonocytic" and "leukemia." The results displayed ("diseases match all terms") will have all three words in the histology name. The words may appear in any part of the entry (alternate names, abstractor notes, transformations, etc.).
        ii. The search engine can also display the number of diseases having at least one of the search words by choosing "diseases match any term"
    c. You can also search on abbreviations such as AMML for acute myelomonocytic leukemia, DLBCL for diffuse large B-cell lymphoma, or AML for acute myeloid leukemia.
    d. When multiple results are displayed, click on the desired term (e.g. acute myelomonocytic leukemia) to display the record.
    e. Search on histology code if desired, i.e., 9867/3.

2. Determine the **number of primaries** using the working histology code(s) with the M rules in the manual

    Note 1: The M rule references in the Heme DB are to be used as a guide only. Start with rule M1, move through the rules in consecutive order and stop at the first rule that applies.

    Note 2: Use the Hematopoietic Multiple Primaries Calculator in the Heme DB only when instructed by the rules in the Hematopoietic Manual.

3. **Verify or revise the working histology code(s)** using the PH rules in the manual (see Note below)
    a. When the PH rules lead you to a different histology code, enter that code in the Heme DB search box and display the record for that histology

    Note: The PH rules referenced in the Heme DB are the most common rule(s) used to code Primary Site and Histology for the selected histology. More than one Module/PH Rule may be needed to code Primary Site and Histology.

4. Determine **primary site** *(see Note on next page)*
    a. See Primary Site Coding Instructions.
    b. For certain histologies, only one primary site code is displayed
        i. The primary site code displayed under **Primary Site(s)** is the **only** site code to be used for that histology
        ii. All leukemia, myelodysplastic syndromes and chronic myeloproliferative diseases are assigned primary site bone marrow C421. There are no exceptions. This rule was implemented in ICD-O-2 in 1992.
    c. When there is no primary site code listed under **Primary Site(s)**
        i. Review the **Primary Site Text** field for common primary sites or other primary site instructions and rules.
        ii. Search the Hematopoietic Manual and/or database to find applicable modules.
        iii. Read the **Abstractor Notes** to find other information regarding sites of involvement for stages II, III, and IV lymphomas. Use the **Abstractor Notes** to confirm that the **site/histology combination indicated by the involvement documented in the medical record is probable**. You may also seek a physician's help in determining the primary site.

(See Note on next page)

***Note*:** Use Modules 1-9 (PH1-PH31) to help determine primary site and histology. Modules 1-6 are histology specific. The remaining are:
Module 7: All lymphomas, extraosseous plasmacytomas, histiocytic and dendritic cell neoplasms, mast cell sarcoma, heavy chain disease, myeloid sarcoma and post-transplant lymphoproliferative disease (PTLD)
Module 8: All hematopoietic neoplasms (NOS and more specified histologies)
Module 9: All hematopoietic neoplasms

5. Determine the **grade**
    a. See the Grade field in the Heme DB
    b. See the Grade rules in the manual when grade cannot be coded using the Heme DB

First Course of Treatment for Hematopoietic Neoplasms

Treatment varies by the type of hematopoietic neoplasm.

Lymphomas can be treated with surgery (extranodal or nodal), chemotherapy and radiation, while leukemias are often treated with chemotherapy and bone marrow transplants. In addition, immunotherapy (biologic response modifiers) and hormones are frequently used to treat hematopoietic neoplasms. Also, for many of these diseases, the principal treatment is either supportive care, observation, or another type of treatment that does not meet the usual definition of treatment that "modifies, controls, removes or destroys proliferating cancer tissue."

Starting in 2010, some neoplasms that have undergone a transformation are reported as new primaries (see rules M10-M13 for specific instructions), and treatment can affect this. For purposes of determining multiple primaries in the Hematopoietic diseases, "treatment" refers to the patient receiving at least one form of cancer-directed treatment such as surgery or systemic therapy, not passive treatment plans like supportive care or observation.

Coding Instructions

1. When there is only one neoplasm (one primary), use the documented first course of therapy (treatment plan) from the medical record. First course of therapy ends when the treatment plan is completed, no matter how long it takes to complete the plan.
2. Chronic neoplasm followed by an acute neoplasm
    a. The presence/absence of treatment **DOES NOT** affect the number of primaries when a chronic neoplasm transforms to an acute neoplasm
    Example*:* Patient diagnosed in 2000 with follicular lymphoma. Patient refused treatment. Patient returns in 2014 with DLBCL. Abstract the DLBCL as a second primary even though there was no treatment for the follicular lymphoma.
    b. First course of treatment for the chronic neoplasm may or may not be completed when the chronic neoplasm transforms to the acute neoplasm.
3. Acute neoplasm followed by a chronic neoplasm
    a. The presence/absence of treatment **DOES** impact the determination of the number of primaries when the acute neoplasm reverts to a chronic neoplasm (see Rules M12 and M13).
    b. The planned first course of therapy may not have been completed when a biopsy/pathologic specimen shows only chronic neoplasm after an initial diagnosis of an acute neoplasm.
    c. The patient may have completed the first course of treatment and have been cancer free (clinically, no evidence of the acute neoplasm) for an interim when diagnosed with the chronic neoplasm.
    d. The patient may not have been cancer free, but completed the first course of treatment and biopsy/pathology shows only chronic neoplasm.

Code the treatment on both abstracts when a patient has multiple primaries and the treatment given for one primary also affects/treats the other primary.

Example: Patient is diagnosed in May 2014 with both multiple myeloma (9732/3) and mantle cell lymphoma (9673/3), which are separate primaries per rule M15. The oncologist states she began Velcade chemotherapy for the lymphoma. Velcade would affect both primaries, so it should be coded on both abstracts.

Reporting Phlebotomy, Blood-Thinners/Anti-Clotting Medications, and Transfusions as Other Therapy (NAACCR Item #1420)

Note: These instructions apply to cases diagnosed 2010+.

- Do **not** collect **blood transfusions** (whole blood, platelets, etc.) as treatment. Blood transfusions are widely used to treat anemia and it is not possible to collect this procedure in a meaningful way.
- Collect **phlebotomy** for polycythemia vera (9950/3) ONLY.
- Collect **blood-thinners** and/or **anti-clotting agents** for
  9740/3 Mast cell sarcoma
  9741/3 Systemic mastocytosis
  9742/3 Mast cell leukemia
  9875/3 Chronic myelogenous leukemia *BCR-ABL1* positive
  9950/3 Polycythemia vera
  9961/3 Primary myelofibrosis
  9962/3 Essential thrombocythemia
  9963/3 Chronic neutrophilic leukemia
  9975/3 Myelodysplastic/myeloproliferative neoplasm, unclassifiable

Donor Leukocyte Infusions

The use of donor leukocyte infusions for treatment of hematopoietic neoplasms, specifically leukemias, is increasing. Abstract as bone marrow transplant when a reportable hematopoietic neoplasm is treated with donor leukocyte infusion, even if it is not listed in the treatment section of the Heme db for the specific neoplasm.

Case Reportability Instructions

Note: Make inquiries to the physician's office to confirm the diagnosis whenever possible. Unless that type of follow-back is done, hematopoietic cases will be under-reported.

1. Search the **Heme DB** to determine case reportability.

2. Report all cases with morphology codes **9590-9992** with a **/3** behavior.
   Note 1: In ICD-O-3 preleukemia is listed as 9989/3 in the numeric list and 9989/1 in the alphabetic list. Change the 9989/1 in the alphabetic list to a 9989/3.
   Note 2: Report the case and change the behavior code to a /0 or /1 in the rare instances of a benign or borderline heme neoplasm occurring in the brain and/or CNS diagnosed 1/1/2004 or later.

3. Report hematopoietic and lymphoid neoplasms with morphology codes **9590-9992** listed in ICD-O-3 as **/1** that are **described as malignant** by a physician. Apply the matrix rule described in ICD-O-3 (Rule F, pg. 29, hardback edition) and change the behavior code to /3.
   Note: Do **not** report in situ (/2) lymphomas.

4. Report the case when the diagnosis of a hematopoietic neoplasm is preceded by one or more of the **ambiguous terms listed below:**
   a. **This instruction pertains to reportability and casefinding only. See the Histology Coding Instructions, #3-5 for instructions on assigning histology with ambiguous terminology.**
      - Apparently
      - Appears
      - Comparable with
      - Compatible with
      - Consistent with
      - Favor(s)
      - Malignant appearing
      - Most likely
      - Presumed
      - Probable
      - Suspect(ed)
      - Suspicious (for)
      - Typical (of)

   Note 1: Use these terms when screening all reports other than cytology and tumor markers.
   Note 2: Report cases that use only 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 with". Do not substitute "likely" for "most likely." See SEER coding manual - Reportability section.
   Note 3: 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 4: Follow back is recommended for diagnoses based on ambiguous terminology to see if the diagnosis has been confirmed or proven to be incorrect (see note 5).
   Note 5: Do **not** report the case when biopsy or physician's statement confirms a non-reportable condition or proves the ambiguous diagnosis is **wrong.**

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 ambiguous diagnosis was wrong. Do **not** report the case.

Note 6: Do **not** report cases diagnosed only by ambiguous **cytology** (cytology diagnosis preceded by ambiguous term).

Example: Parotid ultrasound guided FNA: consistent with non-Hodgkin's lymphoma. This case was diagnosed based on cytology/fine needle aspiration (FNA) preceded by ambiguous terminology (**consistent with**). Do not report this case based on ambiguous cytology.

5. Report the case when the patient is **treated** for a reportable neoplasm.

Note 1: Report the case even if the diagnostic tests are inconclusive, equivocal, or negative.

Note 2: For treatment information see the National Cancer Institute's Physicians' Data Query (PDQ) website at http://www.cancer.gov/cancertopics/pdq or the SEER*Rx Antineoplastic Drugs Database.

6. Report the case when there is a **clinical diagnosis** (physician's statement) of reportable hematopoietic or lymphoid neoplasm.

Note 1: The clinical diagnosis may be a final diagnosis found within the medical record or recorded on a scan (CT, MRI for example).

Note 2: Report the case even if the diagnostic tests are equivocal. A number of hematopoietic neoplasms 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 Heme DB for definitive diagnostic methods for the specific neoplasm being abstracted.

7. Report the case when a reportable diagnosis appears in any text or report described as a **Definitive Diagnostic Method** in the Heme DB.

Note: Definitive diagnostic methods differ depending upon the histology. See the Heme DB for details.

Multiple Primary Rules

General Instructions for Multiple Primary Rules

1. Use the M rule references in the Heme DB as a guide only. Start with M1 for each case, move through the rules and stop at the first rule that applies.
2. Within these rules, the term "chronic neoplasm" means that a neoplasm that has the potential to transform into another, more acute neoplasm. When the chronic neoplasm is displayed in the Heme DB, the "Transformations to" field will show the acute neoplasm.
   Example: Essential thrombocythemia (9962/3). Under the "Transformations to" field, all the acute myeloid leukemias will be listed. In this case, the essential thrombocythemia is the chronic disease while the AML's are the acute diseases.
3. The registrar must recognize that during the diagnosis workup the physician may start with a provisional or several provisional (differential) diagnoses (NOS) and as testing is completed, a more specific diagnosis may be identified. These diagnoses are not multiple primaries, they represent steps in the diagnostic work-up.
4. The Heme DB Multiple Primaries Calculator is to be used only when the rules instruct you to do so.

Rule M1 Abstract 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 a single primary* when there is a single histology.

Note 1: Bilateral involvement of lymph nodes and/or organs with a single histology is a single primary.

Note 2: Recurrence of the same histology is always the same primary (timing is not relevant).

Note 3: A single histology is diagnosed by the definitive diagnostic method as defined in the Heme DB. For example, the patient had several provisional diagnoses but the definitive diagnostic method identified a single histology. Abstract as a single primary.

Example 1: The diagnosis is multiple myeloma (9732/3). Abstract as a single primary.

Example 2: Right and left breast both involved with diffuse large B-cell lymphoma (9680/3). Abstract as a single primary.

Rule M3 Abstract a single primary* when a **sarcoma** is diagnosed simultaneously or after a **leukemia of the same lineage**

- **Mast cell sarcoma (9740/3)** diagnosed simultaneously with or after **mast cell leukemia (9742/3)**
- **Myeloid sarcoma (9930/3)** diagnosed simultaneously with or after **acute myeloid leukemia (9861/3)** or another leukemia of the myeloid lineage **(9840/3, 9865/3-9867/3, 9869/3-9874/3, 9891/3, 9895/3-9898/3, 9910/3, 9911/3 and 9931/3)**
  - ***Exception*: Chronic myeloid leukemia (CML) codes: 9863/3, 9875/3, 9876/3 are not classified as leukemias of the same lineage as myeloid sarcoma**

Note 1: These sarcomas are solid manifestations of the associated leukemias. For example, when acute myeloid leukemia and myeloid sarcoma are diagnosed simultaneously, the myeloid sarcoma is the result of myeloid cells migrating from the bone marrow or blood into tissue. It is part of the disease process for the acute leukemia.

Note 2: See Module 5 (PH9 and PH10) for information regarding primary site and histology

Example: Acute myeloid leukemia (AML) diagnosed in 2012. In 2013, a soft tissue mass was biopsied and the pathology report final diagnosis was myeloid sarcoma. The myeloid sarcoma is a manifestation of the AML. The malignant myeloid cells are present in the blood. One of the malignant myeloid cells lodged in a capillary and grew in the tissue forming a myeloid cell soft tissue mass (referred to as myeloid sarcoma). This is not a second primary; it is a direct result of the myeloid cells circulating in the blood. It is not unlike a solid tumor in the colon metastasizing to the liver.

* Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

Rule M4 Abstract a single primary* when **two or more types of non-Hodgkin lymphoma** are simultaneously present in the **same anatomic location(s)**, such as the same lymph node or lymph node region(s), the same organ(s), and/or the same tissue(s).

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Note 2: Use Rule M15 for simultaneous occurrences of two or more cutaneous lymphomas. Do **not** use this rule for cutaneous lymphomas. Simultaneous occurrences of two or more cutaneous lymphomas, other than an NOS and more specific, are extremely rare. If there are simultaneous cutaneous lymphomas, **DO NOT** use this rule; proceed to **rule M15** (use Multiple Primaries Calculator).

Note 3: When the neoplasm 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 help identifying lymph node names, chains, regions and codes.

Note 4: When the neoplasm is in a more advanced stage, both non-Hodgkin lymphomas may be present in multiple lymph nodes in the same regions as defined by ICD-O-3, or in an organ and that organ's regional lymph nodes, or in multiple organs.

- Although the combination of two or more types of non-Hodgkin lymphoma 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 of two or more types of non-Hodgkin lymphoma, assume that all of the nodes, tissues, and/or organs and associated lymph nodes are involved with the same combination of non-Hodgkin lymphomas.

Note 5: Do **not** query the Heme DB Multiple Primaries Calculator in this situation.

Note 6: See Rules PH11 and PH15 for assigning primary site and histology.

Example: Biopsy of cervical lymph node shows follicular lymphoma and DLBCL. Abstract as a single primary.

Rule M5 Abstract a single primary* when both **Hodgkin and non-Hodgkin lymphoma** are simultaneously present in the **same anatomic location(s)**, such as the same lymph node or same lymph node region(s), the same organ(s), and/or the same tissue(s).

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Note 2: Do **not** query the Heme DB Multiple Primaries Calculator in this situation

Note 3: When the neoplasm 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 help identifying lymph node names, chains, regions and codes.

Note 4: When the neoplasm 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, 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 5: See PH14 for information regarding primary site and histology

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

* Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

Rule M6 Abstract as multiple primaries** when **Hodgkin lymphoma** is diagnosed in one anatomic location and **non-Hodgkin lymphoma** is diagnosed in another anatomic location.

Note: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Example 1: Patient diagnosed with HL in the cervical lymph nodes and with NHL in the GI tract. Abstract as multiple primaries.

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

Example 3: NHL in a right cervical node and HL in a left cervical node. Abstract as multiple primaries. Left and right node chains are separate regions. See Appendix C.

Rule M7 Abstract as a single primary* when a more specific histology is diagnosed after an NOS **ONLY** when the Heme DB Multiple Primaries Calculator confirms that the NOS and the more specific histology are the same primary.

Note 1: The more specific histology confirmation does not have to occur in the same anatomic location.

Note 2: There are no time restrictions on these diagnoses; the interval between the NOS and the more specific histology does not affect this rule.

Note 3: The Heme DB Multiple Primaries Calculator will identify these histologies as a single primary.

Note 4: Change the histology code on the original abstract to the more specific histology when the original diagnosis is in your registry database. Use previous editions of ICD-O (i.e. ICD-O-1, ICD-O-2) or the Heme DB to assign the code applicable to the year of diagnosis for the more specific histology. .

Example 1: Patient diagnosed with non-Hodgkin lymphoma (9591/3) in 2003. Patient returns in 2013 with a diagnosis of CD30 positive lymphoproliferative disorder (9718/3). 9591/3 is an NOS histology and 9718/3 is more specific. Per the Multiple Primaries Calculator, 9591/3 and 9718/3 are the same primary. 9718/3 was a valid code in 2003; change the histology to 9718/3 for the 2003 diagnosis.

Example 2: CT guided core biopsy pelvic mass positive for lymphoma (9590/3) diagnosed in 2008. In November 2014, Mediastinoscopy with biopsy shows intravascular large B-cell lymphoma. (9712/3). 9590/3 is an NOS histology and 9712/3 is more specific. Per the Multiple Primaries Calculator, 9590/3 and 9712/3 are the same primary. Per the Hematopoietic Database, 9712/3 was not valid until 2010. Since the original diagnosis was in 2008, 9712/3 cannot be used. Keep the original code of 9590/3.

Rule M8 Abstract as a single primary* and code the acute neoplasm when both a **chronic** and an **acute** neoplasm are diagnosed **simultaneously or within 21 days AND** there is documentation of **only one** positive biopsy (bone marrow biopsy, lymph node biopsy, or tissue biopsy).

Note 1: When these diagnoses happen within 21 days, it is most likely that one diagnosis was provisional and the biopsy identified the correct diagnosis. Abstract the acute neoplasm.

Note 2: "Transformations to" (acute neoplasms) and "Transformations from" (chronic neoplasms) are defined for each applicable histology in the database.

Example: Clinical workup shows plasmacytoma (9731/3). Lytic lesions also seen on clinical workup. Bone marrow biopsy done which shows multiple myeloma. Plasmacytoma transforms to multiple myeloma. Code the multiple myeloma (9732/3) since this is the acute neoplasm and there is only one bone marrow biopsy.

Rule M9 Abstract a single primary* and **code the later diagnosis** when both a **chronic** and an **acute** neoplasm are diagnosed **simultaneously or within 21 days AND** there is **no available documentation** on biopsy (bone marrow biopsy, lymph node biopsy, or tissue biopsy.) The later diagnosis could be either the chronic or the acute neoplasm.

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 and correlated with any clinical information.

Note 2: "Transformations to" (acute neoplasms) and "Transformations from" (chronic neoplasms) are defined for each applicable histology in the database.

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* 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.

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

Note 1: **This is a change from the pre-2010 rules**. Use the Heme DB Multiple Primaries Calculator to determine multiple primaries when a transformation from a chronic to an acute neoplasm occurs.

Note 2: "Transformations to" (acute neoplasms) and "Transformations from" (chronic neoplasms) are defined for each applicable histology in the database.

Example: Patient was diagnosed with MDS, unclassifiable in 2010. The patient presents in 2013 with a diagnosis of acute myeloid leukemia (AML) (9861/3). The transformation paragraph in the Heme DB says MDS (chronic neoplasm) transforms to AML (acute neoplasm). Because the chronic neoplasm (MDS) and the acute neoplasm (AML) are diagnosed more than 21 days apart, abstract the MDS and the AML (9861/3) as multiple primaries.

Rule M11 Abstract as multiple primaries[**] when both a **chronic** and an **acute** neoplasm are diagnosed **simultaneously or within 21 days AND** there is **documentation of two** bone marrow examinations, lymph node biopsies, or tissue biopsies: one confirming the **chronic** neoplasm and another confirming the **acute** neoplasm.

Note: "Transformations to" (acute neoplasms) and "Transformations from" (chronic neoplasms) are defined for each applicable histology in the database.

Example: Vertebral biopsy on 2/13/2013 positive for plasmacytoma and 3/2/2013 bone marrow biopsy was positive for multiple myeloma. Biopsies and diagnoses were less than 21 days apart. Code as two primaries, solitary plasmacytoma of bone (9731/3) and plasma cell myeloma/multiple myeloma (9732/3).

Rule M12 Abstract a single primary[*] when a neoplasm is **originally diagnosed as acute AND reverts** to a **chronic** neoplasm **AND** there is no confirmation available that the patient has been treated for the acute neoplasm.

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

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

Note 3: "Transformations to" (acute neoplasms) and "Transformations from" (chronic neoplasms) are defined for each applicable histology in the database.

Example: 3/16/2013 biopsy of cervical nodes positive for diffuse large B-cell lymphoma (DLBCL) (9680/3). 4/18/2013 bone marrow shows follicular lymphoma (9690/3). No treatment given between the diagnoses of acute neoplasm (DLBCL) and chronic (follicular). Abstract one primary, DLBCL (9680/3).

Rule M13 Abstract multiple primaries[**] when a neoplasm is **originally** diagnosed as **acute AND reverts** to a **chronic** neoplasm **after treatment**.

Note 1: Only abstract as multiple primaries when the patient has been treated for the acute neoplasm.

Example: Patient was diagnosed in 2009 with AML, NOS (9861/3). The patient was treated with chemotherapy and a subsequent stem cell transplant. On 2/25/2013 a bone marrow biopsy was positive for myelodysplastic syndrome. Abstract a second primary with the histology MDS (9989/3).

Note 2: Apply this rule when treatment for the acute neoplasm is given, even when all planned treatment is not completed.

Example: Patient diagnosed with AML (9861/3). Plan of treatment chemotherapy. If remission achieved, followed by bone marrow transplant. After chemotherapy, bone marrow biopsy is done and shows a complete remission regarding the AML, but the bone marrow shows MDS (9989/3). The MDS is a second primary even though the planned first course of treatment was not completed prior to the diagnosis of the MDS.

Note 3: The rules regarding first course of treatment are not the same for Solid Tumors and Hematopoietic. Do not apply the Note 2 to Solid Tumors.

Note 4: "Transformations to" (acute neoplasms) and "Transformations from" (chronic neoplasms) are defined for each applicable histology in the database.

[*] 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.

Rule M14 Abstract a single primary* when post-transplant lymphoproliferative disorder is diagnosed **simultaneously** with any B-cell lymphoma, T-cell lymphoma, Hodgkin lymphoma or plasmacytoma/myeloma.
Note 1: **This is a change from previous instructions.** Previously, lymphomas were listed as PTLD transformations. If there is a diagnosis of a lymphoma **AFTER** PTLD, abstract it is a second primary.
Note 2: See Rule PH1 for information regarding histology and Module 7 for assigning primary site.
Note 3: Registrars are NOT required to review cases previously abstracted.

Rule M15 Use the Heme DB Multiple Primaries Calculator to determine the number of primaries for all cases that do not meet the criteria of M1-M14.
Example: Polycythemia vera (PV) diagnosed in 2001, receiving anagrelide. Increasing leukocytosis seen, bone marrow biopsy done in 2013 showing primary myelofibrosis (PMF) with myeloid metaplasia. No rule in M1-M14 applies. Abstract multiple primaries because the Multiple Primaries Calculator shows that PV (9950/3) and PMF (9961/3) are separate primaries.

This is the end of the rules for determining the number of primaries.

* Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes.

Primary Site Coding Instructions

Instructions for assigning primary site have been added to this manual. These instructions correspond to the PH rules and to information in the Heme DB and apply to cases diagnosed 2010 and forward. The primary site code assignments have not changed.

1. Use these instructions, the PH Rules, and the Heme DB to code primary site.
2. Refer to two fields in the Heme DB for primary site instructions:
    a. Primary site: When applicable, a specific site (topography) code(s) will be listed.
    b. Primary site text: This field, new in 2015, provides additional information on assigning primary site. Information on common primary sites (when applicable) has been moved from the Abstractor Notes to this field.
    Example 1*:* Histology 9800/3. Primary site field has "C421." Primary site text field has "Primary site must be bone marrow (C421)." For this histology, the primary site is C421 for all cases with this histology. There are no exceptions.
    Example 2**:** Histology 9650/3. Primary site field has "C770-C779." Primary site text field has "Lymph nodes (C770-C779) are the usual primary sites; however, involvement in other sites is possible. If you have confirmation that the only site is something other than the lymph nodes, then code to that primary site. See also Module 7." For this histology, the preferred primary sites are the lymph nodes; however, other sites are possible, but rare.

3. Code **primary site** using
    - Scans
    - Medical record documentation
    - Pathology report
    - Heme DB

    Note 1: Do not simply code the site of a lymph node biopsy; use the information available from scan to determine the correct primary site.
    Note 2: There is no hierarchy among the items on this list.
    Note 3: For hematopoietic neoplasms, the pathology report is not the default for determining the primary site, especially for lymphoma. The standard for determining primary site differs depending upon the specific histology.
    Note 4: If a hematopoietic neoplasm is diagnosed by peripheral blood smear and no other information is available, assign the primary site as bone marrow (C421). (See Rule PH26, Note 2).

4. Code the primary site as indicated below for the following histologies.
    Note 1: Use of either the Heme DB primary site coding instructions or the instructions below will result in the same primary site code.
    Note 2: Do not assign primary site codes C423 (Reticuloendothelial system, NOS) or C424 (Hematopoietic system, NOS) for the Hematopoietic neoplasms. CSv0205 allows these primary sites; however, revised edits will not allow them.

    A. Primary site C379 (Thymus) or C383 (Anterior Mediastinum). Assign primary site to C379 or C383 when the histology is:

    9679/3-Primary mediastinal (thymic) large B-cell lymphoma
    Note: **Do not code this histology** based only on mediastinal involvement. Only assign this histology code when the diagnosis is stated as "primary mediastinal" large B-cell lymphoma.

    B. Primary site C400-C419 (Bone). The following histology is always coded to primary site C400-C419:

    9731/3-Solitary plasmacytoma of bone
    Note: 9731/3 for plasmacytomas of the bone. If there is an extramedullary plasmacytoma (not occurring in bone) see histology 9734/3. (See also PH3 & PH4).

C. Primary site C420 (Blood). Assign primary site to C420 (Blood) when the histology is

9761/3-Waldenstrom Macroglobulinemia (9761/3) (See also Rules PH16 & PH17)
Note: Do not code primary site of blood for any other hematopoietic histology (9590/3-9992/3)

D. Primary site C421 (Bone marrow). Assign primary site C421 (Bone marrow) when the histology is:

9732/3-Plasma cell myeloma
9741/3-Systemic mastocytosis
9742/3-Mast cell leukemia
9800/3-Leukemia, NOS
9801/3-Acute leukemia, NOS
9806/3-Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1
9807/3-Mixed phenotype acute leukemia with t(v;11q23); MLL rearranged
9808/3-Mixed phenotype acute leukemia, B/myeloid, NOS
9809/3-Mixed phenotype acute leukemia, T/myeloid, NOS
9820/3-Lymphoid leukemia, NOS
9826/3-Burkitt cell leukemia
9831/3-T-cell large granular lymphocytic leukemia
9832-3-Prolymphocytic leukemia, NOS
9833/3-B-cell prolymphocytic leukemia
9834/3-T-cell prolymphocytic leukemia
9840/3-Acute erythroid leukemia
9860/3-Myeloid leukemia
9861/3-Acute myeloid leukemia, NOS
9863/3-Chronic myeloid leukemia
9865/3-Acute myeloid leukemia with t(6;9)(p23;q34) *DEK-NUP214*
9866/3-Acute promyelocytic leukemia with t(15;17)(q22;q12), *PML-RARA*
9867/3-Acute myelomonocytic leukemia
9869/3-Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26.2); *RPN1-EVI1*
9870/3-Acute basophilic leukemia
9871/3-Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); *CBFB-MYH11*
9872/3-Acute myeloid leukemia with minimal differentiation
9873/3-Acute myeloid leukemia without maturation
9874/3-Acute myeloid leukemia with maturation
9875/3-Chronic myelogenous leukemia, *BCR-ABL1* positive
9876/3-Atypical chronic myeloid leukemia, *BCR-ABL1* negative
9891/3-Acute monoblastic and monocytic leukemia
9895/3-Acute myeloid leukemia with myelodysplasia-related changes
9896/3-Acute myeloid leukemia with myelodysplasia-related changes
(list continued on next page)

9897/3-Acute myeloid leukemia with t(9;11)(p22;q23); *MLLT3-MLL*
9898/3-Myeloid leukemia associated with Down syndrome
9910/3-Acute megakaryoblastic leukemia
9911/3-Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); *RBM15-MKL1*
9920/3-Therapy-related myeloid neoplasm
9931/3-Acute panmyelosis with myelofibrosis
9940/3-Hairy cell leukemia
9945/3-Chronic myelomonocytic leukemia
9946/3-Juvenile myelomonocytic leukemia
9948/3-Aggressive NK-cell leukemia
9950/3-Polycythemia vera
9961/3-Primary myelofibrosis
9962/3-Essential thrombocythemia
9963/3-Chronic neutrophilic leukemia
9964/3-Chronic eosinophilic leukemia, NOS
9965/3-Myeloid and lymphoid neoplasms with PDGFRA rearrangement
9966/3-Myeloid neoplasm with PDGFRB rearrangement
9967/3-Myeloid and lymphoid neoplasm with FGFR1 abnormalities
9975/3-Myelodysplastic/myeloproliferative neoplasm, unclassifiable
9980/3-Refractory anemia
9982/3-Refractory anemia with ring sideroblasts
9983/3-Refractory anemia with excess blasts
9985/3-Refractory cytopenia with multilineage dysplasia
9986/3-Myelodysplastic syndrome associated with isolated del (5q)
9989/3-Myelodysplastic syndrome, unclassifiable
9991/3-Refractory neutropenia
9992/3-Refractory thrombocytopenia

E. Primary site C422 (Spleen). Assign primary site C422 (Spleen) when the histology is:

9689/3-Splenic marginal zone lymphoma
9716/3-Hepatosplenic T-cell lymphoma
Note: Other lymphomas (e.g. DLBCL), may originate in the spleen, although this is rare. Spleen involvement does not necessarily mean the primary site is the spleen.

F. Primary site C440-C449, C510-C512, C518-C519, C600-C602, C608-C609, C632 (Skin primary sites) should be assigned when the histology is:

***Note*:** The following histologies are skin lymphomas.
9597/3-Primary cutaneous follicle centre lymphomas (See also Rule PH12)
9700/3-Mycosis fungoides
9701/3-Sezary syndrome
9708/3-Subcutaneous panniculitis-like T-cell lymphoma (also C490-C499)
9709/3-Primary cutaneous T-cell lymphoma
9718/3-Primary cutaneous anaplastic large cell lymphoma
9725/3-Hydroa vacciniforme-like lymphoma
9726/3-Primary cutaneous gamma-delta T-cell lymphoma (also C490-C499)

G. Primary sites C770-C779 (Lymph nodes) should be assigned **unless** there is confirmation that the primary site is extranodal when the histology is:

***Note*:** The following histologies usually originate in the lymph nodes; however, in rare cases they may originate in extranodal sites
9596/3-B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma (See also Rule PH14)
9650/3-Classical Hodgkin lymphoma
9651/3-Lymphocyte-rich classical Hodgkin lymphoma
9652/3-Mixed cellularity classical Hodgkin lymphoma
9653/3-Lymphocyte-depleted classical Hodgkin lymphoma
9655/3-Hodgkin lymphoma, lymphocyte depletion, reticular
9659/3-Nodular lymphocyte predominant Hodgkin lymphoma
9663/3-Nodular sclerosis classical Hodgkin lymphoma
9688/3-T-cell/histiocyte-rich large B-cell lymphoma

H. Assign the primary site code using **Modules 3 & 4: Rules PH5-PH8** when the histology is**:**

Note 1: These are lymphoma/leukemias and primary site code is based on presentation
Note 2: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.
- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

9727/3-Blastic plasmacytoid dendritic cell neoplasm (Blastic natural killer leukemia/lymphoma)
9811/3-B lymphoblastic leukemia/lymphoma, NOS
9812/3-B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); *BCR-ABL1*
9813/3-Lymphoblastic leukemia/lymphoma with t(v;11q23); *MLL* rearranged
9814/3-B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); *TEL-AML1 (ETV6-RUNX1*)
9815/3-B lymphoblastic leukemia/lymphoma with hyperdiploidy
9816/3-B lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL)
9817/3-B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); *IL3-*
9818/3-B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); *E2A-PBX1 (TCF3-PBX1)*
(List continued on next page)

9823/3-Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL)
9827/3-Adult T-cell leukemia/lymphoma (HTLV-1 positive
9837/3-Adult T-cell leukemia/lymphoma (HTLV-1 positive)

I. Assign the primary site code using Module 7 for lymphomas, extraosseous plasmacytomas, mast cell sarcoma, histiocytic and dendritic cell neoplasms, heavy chain disease, myeloid sarcoma and PTLD

***Note*:** More than one PH rule will be required to code the primary site for some cases.

Example 1: Patient diagnosed with CLL/SLL in the cervical lymph nodes. Per Rule PH6 (See Rule G), code primary site to the involved site. Per Note 5, go to Module 7 to code primary site for the lymphoma. Per Rule PH19, code the primary site to the specific lymph node region when only one lymph node is involved. Code primary site to C771, cervical lymph node.

Example 2: Lymph node biopsy confirmed plasmacytoma. Adenopathy in multiple locations. Per Rule PH2, code the histology to 9734/3 and the primary site to the involved site. Per Note 6, go to Module 7 to code primary site for the lymphoma. Per Rule PH22, code the primary site to lymph nodes, NOS (C779) when multiple lymph nodes are involved but there is no particular lymph node region identified.

9590/3-Malignant lymphoma, NOS
9591/3-Non-Hodgkin lymphoma, NOS
9671/3-Lymphoplasmacytic lymphoma (See also Rules PH16 & PH17)
9673/3-Mantle cell lymphoma
9678/3-Primary effusion lymphoma
9680/3-Diffuse large B-cell lymphoma (DLBCL) (See also Rule PH12)
9687/3-Burkitt lymphoma
9690/3-Follicular lymphoma, NOS
9691/3-Follicular lymphoma, grade 2
9695/3-Follicular lymphoma, grade 1
9698/3-Follicular lymphoma, grade 3
9699/3-Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)
9702/3-Peripheral T-cell lymphoma, NOS
9705/3-Angioimmunoblastic T-cell lymphoma
9712/3-Intravascular large B-cell lymphoma
9714/3-Anaplastic large cell lymphoma, ALK-positive
9717/3-Enteropathy-associated T-cell lymphoma
9719/3-Extranodal NK-/T-cell lymphoma, nasal type
9724/3-Systemic EBV-positive T-cell lymphoproliferative disease of childhood
9734/3-Extraosseous Plasmacytoma (See also rule PH2)
9735/3-Plasmablastic lymphoma
9737/3-ALK-positive large B-cell lymphoma
9738/3-Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease
(List continued on next page)

9740/3-Mast cell sarcoma (See also Rules M3 & PH9)
9751/3-Langerhans cell histiocytosis
9755/3-Histiocytic sarcoma
9756/3-Langerhans cell sarcoma
(list continued on next page)
9757/3-Interdigitating dendritic cell sarcoma
9758/3-Follicular dendritic cell sarcoma
9759/3-Fibroblastic reticular cell tumor
9762/3-Heavy chain disease
9930/3-Myeloid sarcoma (See also Rules M3 & PH10)
9971/3-Polymorphic post-transplant lymphoproliferative disorder (PTLD) (See also Rule PH1)

Histology Coding Instructions

1. Code the **histology** that was identified by the method(s) listed under the **Definitive Diagnostic Method(s)** section of the Heme DB. Definitive diagnostic method(s) may be any of the following
   Note: There is no hierarchy among the items on this list
   - Clinical diagnosis
   - Genetic test
   - Immunophenotyping
   - Cytology
   - Pathology
     - Final diagnosis
     - Comment on final diagnosis
     - Addenda to final diagnosis
     - CAP protocol/synoptic report

2. When tests or reports defined as Definitive Diagnostic Method(s) are **not available**, code histology using the following documentation. The list is in **hierarchical order.**
   - **Documentation** in the medical record referring to the **original** scans, genetic testing, immunophenotyping, or pathology reports
   - **Documentation** in the medical record that refers to the histology
   - Death certificate (central or regional registries only)

3. When the test or report lists a specific histology with **ambiguous term(s)** and an "NOS" histology, code the **NOS histology**. This prevents coding a temporary/provisional histology that could change with further testing which may not appear in the patient's chart, such as subsequent flow cytometry sent from the physician's office to an outside lab. (See also Rule PH29)
   Note 1: Ambiguous terminology can be used for casefinding, reportability, and to assign a provisional histology code. For instruction on using ambiguous terminology for casefinding and reportability, go to the "Case Reportability Instructions, #4" at the beginning of the manual.
   Note 2: For hematopoietic and lymphoid neoplasms, ambiguous terminology may be used when a specific histology has not been confirmed. In this situation, there is not enough proof for the physician to definitely diagnose the specific histology. If there is no further information regarding the more specific histology, the registrar is to assign the NOS equivalent for that histology.
   Example 1: Non-Hodgkin lymphoma consistent with DLBCL. No other information available. "Non-Hodgkin Lymphoma" is listed as an "NOS" histology in the Heme DB. Because it is preceded by the ambiguous term "consistent with", the more specific histology DLBCL would not be coded until confirmed by further testing. Assign 9591/3 for Non-Hodgkin lymphoma, NOS.
   Note 3: A specific histology can be assigned if there is documentation that the physician is treating the patient for the specific disease.
   Example 2: Biopsy: B-cell lymphoma, suspicious for DLBLC. Subsequent documentation from the physician indicates that the patient is being treated for DLBCL.

4. If there is only one histology available and it is preceded by ambiguous terminology, review the Abstractor Notes in the Heme DB for that histology to see if other information can be used to confirm the diagnosis.
   Example: CBC states abnormal lymphocytosis, no histology or provisional diagnosis on the CBC or peripheral blood smear. Flow cytometry states compatible with CLL. No other workup done. Per the Abstractor Notes in the database, "abnormal lymphocytosis" is present in CLL. Assign histology CLL (9823/3) since "abnormal lymphocytosis" is part of the CLL/SLL definition.

5. If the Abstractor Notes, immunophenotyping or genetics information and the only histology available is preceded by ambiguous terminology, code the ambiguous histology so that the case can be reported for incidence. (See Case Reportability Instructions, #4). Follow back to the physician's office or other sources at a later date to determine if subsequent testing confirmed the specific histology or the patient was diagnosed with a different histology.

Primary Site and Histology Coding Rules

1. The primary site and histology coding rules are divided into nine modules. The first six modules cover primary site and histology; the last three cover coding primary site only. Each **module** covers a group of **related** hematopoietic or lymphoid neoplasms. However, a specific histology may be covered in more than one module.
2. 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.

Module 1: Post-Transplant Lymphoproliferative Disorder PH1

Post-transplant lymphoproliferative disorder with accompanying lymphoma or plasmacytoma/myeloma

Rule PH1 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), or organ(s), and code the **histology of the accompanying lymphoma or plasmacytoma/myeloma when the diagnoses of post-transplant lymphoproliferative disorder and any B-cell lymphoma, T-cell lymphoma, Hodgkin lymphoma, or plasmacytoma/myeloma occur simultaneously.**

Note 1: Rule PH1 applies to monomorphic post-transplant lymphoproliferative disorders. The diagnosis may or may not include the word "monomorphic".
Note 2: For **polymorphic PTLD** (PTLD by itself**),** see Rule PH30 and the Heme DB (9971/3).
Note 3: The patient **must** have a history of a solid organ transplant or an allogeneic bone marrow transplant.
Note 4: Most cases of PTLD occur within a year of transplantation; however, they can occur any time after the transplant.
Note 5: Monomorphic PTLD is also caused by the immunosuppressant drugs. Patients are treated for the lymphoma or plasmacytoma/myeloma.
Note 6: See Appendix C for help identifying lymph node names, chains, regions and codes.
Note 7: See Module 7 **for help in coding primary site for PTLD.**
Example: Previous history of kidney transplant. Now presents for bone marrow biopsy. BM positive for B-cell lymphoma. Abdominal mass biopsy was positive for PTLD, monomorphic type and aggressive B-cell malignancy. Immunohistochemistry shows the B-cell malignancy to be Burkitt lymphoma. Code the histology to Burkitt lymphoma and primary site to the abdominal lymph nodes (C77.2).

Module 2: Plasmacytomas PH2 – PH4

Extraosseous plasmacytoma (9734/3)
Solitary plasmacytoma of bone (9731/3)

Note: For information on Plasma Cell Myeloma/Multiple Myeloma (9732/3), see the Hematopoietic database.

Rule PH2 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), or organ(s), and code the **histology extramedullary plasmacytoma** (**9734/3)** when any of the following occur in a site **other than bone:**

- Extraosseous (extramedullary) plasmacytoma
- Multiple extraosseous (extramedullary) plasmacytomas
- Multiple plasmacytomas
- Plasmacytoma, NOS (not occurring in bone)
- Solitary plasmacytoma

Note 1: Extramedullary and extraosseous mean outside of 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 the primary site to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423) or the hematopoietic system, NOS (C424).
Note 4: See Appendix C for help identifying lymph node names, chains, regions and codes.
Note 5: See Module 7 for help in coding primary site for extraosseous plasmacytomas.
Example 1: Pathology reports a solitary plasmacytoma wrapped around L4 vertebrae, no invasion of vertebrae. Code the primary site as soft tissue of back (C496) and histology to plasmacytoma (9734/3).
Example 2: Scan shows two plasmacytomas in the nasopharyngeal wall. Biopsy confirms plasmacytoma. Code the primary site nasopharynx (C119) and histology to plasmacytoma (9734/3).

Rule PH3 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:

- Multiple medullary plasmacytomas
- Multiple plasmacytomas
- Multiple plasmacytomas of bone
- Plasmacytoma, NOS (occurring in bone)
- Solitary medullary plasmacytoma
- Solitary plasmacytoma
- Solitary plasmacytoma of bone

Note 1: **Plasma cell neoplasm** is not an alternate name for 9731/3 and has been **removed** from the alternate names list for 9731/3.
Note 2: 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 3: When multiple bone sites are involved that are not included in the same ICD-O-3 code, code primary site to bone, NOS (C419).
Note 4: Do **not** code primary site to blood (C420), bone marrow (C421), reticuloendothelial system, NOS (C423), or the hematopoietic system, NOS (C424).

Rule PH4 Code the primary site **bone, NOS** (**C419)** and histology **solitary plasmacytoma, NOS (9731/3)** when the only information is that the patient had a **plasmacytoma, NOS** or a **solitary plasmacytoma**, **NOS** and there is no indication of bone or extramedullary.

Note: Default to coding plasmacytoma of bone when the only information available is that the patient had a plasmacytoma (See 9731/3 in the Heme DB).

Example: Death-certificate-only case (central or regional registry only) with underlying cause of death listed as plasmacytoma.

Module 3: Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL) PH5-PH6

Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL) 9823/3

Rule PH5 Code the **primary site** to bone marrow (C421) when the bone marrow is involved or when only peripheral blood is involved.

Note 1: Do **not** code primary site to blood (C420) even when CLL/SLL is diagnosed only on peripheral blood. CLL/SLL will always have peripheral blood involvement. The bone marrow may or may not be involved. Assign primary site to C421 (bone marrow).

Example: Positive peripheral smear for CLL with clinical lymph node involvement. No bone marrow biopsy done. Code primary site to C421 since the peripheral blood is involved.

Note 2: In the later stages of CLL/SLL, there may be involvement of bone marrow AND lymph node(s), lymph node region(s), organ(s), or tissue(s). As long as the peripheral blood and/or bone marrow are involved, the primary site is bone marrow (C421). If **peripheral blood and bone marrow are not involved**, **see Rule PH6.**

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

Rule PH6 Code the **primary site** to the **involved lymph node(s) or lymph node region(s), the involved organ(s), or tissue(s)** when there is **no peripheral blood involvement AN**D **no bone marrow involvement or when it is unknown if bone marrow is involved**.

Note 1: If **peripheral blood and/or bone marrow are involved, see Rule PH5.**

Note 2: CLL/SLL will always have involvement of lymph node(s) or lymph node region(s), organ(s) or tissue(s).

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

Note 4: See Appendix C for help identifying lymph node names, chains, regions and codes.

Note 5: See Module 7 for help in coding primary site for lymphomas.

Module 4: Lymphoma/Leukemia (Specific neoplasms that can manifest as either leukemia or lymphoma or both leukemia and lymphoma) PH7 – PH8 (9727/3, 9811/3-9818/3, 9827/3, 9837/3)

Adult T-cell leukemia/lymphoma (HTLV-1 positive) (9827/3)
Adult T-cell leukemia/lymphoma (9837/3)
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(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(12;21)(p13;q22); *TEL-AML1 (ETV6-RUNX1*) (9814/3)
B lymphoblastic leukemia/lymphomas, NOS (9811/3, 9812/3-9818/3)
Blastic plasmacytoid dendritic cell neoplasm (Blastic natural killer leukemia/lymphoma) (9727/3)
Lymphoblastic leukemia/lymphoma with t(v;11q23); *MLL* rearranged (9813/3)

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.
- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Note 2: ICD-9-CM, ICD-10, and ICD-10-CM have separate codes for leukemia and lymphoma.
Note 3: Lymphoma commonly originates in lymph nodes, tissue, or an organ although it will metastasize to the bone marrow when the stage is IV or disseminated.
Note 4: Liver is usually a metastatic site; however, primary liver lymphoma is possible.

Rule PH7 For the histologies listed above, code the primary site to bone marrow (C421) when the **only** site involved is bone marrow.
Note 1: **If lymph node(s), lymph node region(s), organ(s) or tissue(s) are involved, see Rule PH8.**
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 PH8 For the histologies listed above, code the primary site to the site of origin when lymph node(s) or lymph node region(s), tissue(s) or organs are involved.
Note 1: Do **not** simply code the site of a biopsy; also use the information available from scans to determine the correct primary site.
Note 2: Bone marrow may or may not be involved. If bone marrow is involved, code this information in CS Extension.
Note 3: See Appendix C for help identifying lymph node names, chains, regions and codes.
Note 4: See Module 7 for more information on coding primary site for lymphomas.

Module 5: Myeloid Neoplasms and Mast Cell Neoplasms PH9 - PH10

Myeloid leukemias: (9840/3, 9861/3, 9865/3-9867/3, 9869/3-9874/3, 9891/3, 9895/3-9898/3, 9910/3, 9911/3, 9931/3)
Mast cell leukemia (9742/3)
Mast cell sarcoma (9740/3)
Myeloid sarcoma (9930/3)

Rule PH9 Code the primary site **bone marrow (C421)** and code the **histology** to **mast cell leukemia** (**9742/3)** when the diagnosis is **mast cell sarcoma AND** there is a simultaneous or previous diagnosis of **mast cell leukemia.** See Rule M3.

Note: When mast cell sarcoma (9740/3) follows a diagnosis of mast cell leukemia (9742/3), the sarcoma is a manifestation of late-stage leukemia. The mast cells infiltrate soft tissue.

Rule PH10 Code the primary site **bone marrow (C421)** and code the histology **acute myeloid leukemia, NOS (9861/3) or any of the specific AML histologies (9840/3, 9865/3-9867/3, 9869/3-9874/3, 9891/3, 9895/3-9898/3, 9910/3, 9911/3 and 9931/3)** when the diagnosis is **myeloid sarcoma** (9930/3) **AND** there is a simultaneous or previous diagnosis of acute myeloid leukemia. See Rule M3.

Note: When myeloid sarcoma (9930/3) follows a diagnosis of acute myeloid leukemia (histologies listed above), the sarcoma is a manifestation of late-stage leukemia.

Module 6: Coding Primary Site for Specified Lymphomas PH11 – PH17

Non-Hodgkin lymphomas (9590/3, 9591/3, 9670/3-9679/3, 9684/3-9729/3, 9735/3-9738/3, 9811/3-9818/3, 9823/3, 9827/3, 9837/3)
B-cell lymphoma, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma (9596/3)
Diffuse large B-cell lymphoma (9680/3)
Lymphoplasmacytic lymphoma (9671/3)
Primary cutaneous follicle centre lymphoma (9597/3)
Waldenstrom macroglobulinemia (9761/3)

Note 1: Liver is usually a metastatic site; however, primary liver lymphoma is possible.
Note 2: Do not simply code the site of a biopsy; use the information available from scans to determine the correct primary site.
Note 3: See Primary Site Coding Instructions and Module 7 for more information on coding primary site for lymphoma.

Rule PH11 Code the primary site to the **site of origin**, lymph node(s), lymph node region(s), tissue(s) or organ(s) **and histology** to **diffuse large B-cell lymphoma** (DLBCL) (9680/3) when DLBCL and any other B-cell non-Hodgkin lymphoma are present in the same lymph node(s), lymph node region(s), organ(s), tissue(s) or bone marrow. See Rule M4.

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Note 2: Use this rule when the diagnosis of DLBCL and another non-Hodgkin lymphoma are **diagnosed simultaneously in the same location.**

Note 3: Go to Rule PH15 if one or more of the histologies are not a B-cell.
Note 4: See Appendix C for help identifying lymph node names, chains, regions and codes.
Note 5: See Module 7 for more information on coding primary site for lymphomas.

Rule PH12 Code the primary site to **skin** (C44_) and the histology to **primary cutaneous follicle center 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 node(s)**

Note 1: All variants of follicular lymphoma (NOS, grade 1, grade 2, and grade 3) were once called "follicle center lymphoma." Although that term is obsolete, it is sometimes used to describe follicular lymphoma. You may also see "follicle center" in the pathology reports for follicular lymphoma. However, the primary site and other sites of involvement will differ between follicular lymphoma and follicle center lymphoma. Follicle center lymphoma is a cutaneous malignancy with only rare involvement of regional lymph nodes. Follicular lymphoma commonly occurs in nodes and extranodal sites. (See the Heme DB Abstractor Notes for both neoplasms for information on clinical presentation and common primary sites.)

Note 2: If there is involvement of lymph node(s) 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. Code histology to follicular lymphoma (9690/3, 9691/3, 9695/3 or 9698/3). See the Heme DB for information on coding follicular lymphoma.

Rule PH13 Code the primary site to **skin** (C44_) and the histology to **diffuse large B-cell lymphoma** (9680/3) when there is **skin infiltration** with **large B-cell lymphoma, B-cell lymphoma**, **large cell type, or large cell lymphoma** and the involvement is:

- Limited to **skin or**
- Limited to **skin** and the **regional lymph node(s)**

Note: If there is involvement of lymph node(s) 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 PH14 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow and the histology B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma/**composite Hodgkin and non-Hodgkin lymphoma (9596/3)** when **both** non-Hodgkin lymphoma and Hodgkin lymphoma are **simultaneously** present in the **same** lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow. See Rule M5.

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.
- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Note 2: 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 **or**
- Both NHL and HL are present in multiple lymph node regions as defined by ICD-O-3. e.g., NHL and HL present in superior hilum and superior rectal lymph nodes.
  - When only one node is biopsied, assume all lymph nodes are involved with both NHL and HL.

Note 3: See Appendix C for help identifying lymph node names, chains, regions and codes.
Note 4: See Module 7 for more information on coding primary site for lymphoma.
Note 5: Do **not** use the composite lymphoma histology code (9596/3) when
- NHL is present in one node and HL in another node within the same chain, e.g. NHL in one cervical lymph node and HL in another cervical lymph node **or**

- NHL is present in one lymph node region and HL is present in another lymph node region, e.g., NHL in cervical lymph node(s) and HL in inguinal lymph node(s) **or**
- NHL and HL in different tissues, e.g., NHL in liver and HL in intra-thoracic lymph nodes

Rule PH15 Code the primary site to the **site of origin**, lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow and code the histology to the NHL with the **numerically highest ICD-O-3 code** when two or more **non-Hodgkin lymphomas** are present in the **same** lymph node(s) or lymph node region(s), tissue(s), organ(s), or bone marrow. See Rule M4.

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

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

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

Note 4: This rule does **not** apply to an NOS and more specific histology.

Note 5: This rule does **not** apply when different NHLs are 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.

Example: Biopsy revealed both small lymphocytic lymphoma (9823/3) and follicular lymphoma grade 2 (9691/3) in the same lymph node. Per Rule M4, this is one primary. Code histology to 9823/3 since it is the numerically higher ICD-O-3 code.

Rule PH16 Code the primary site **blood** (C420) and the histology **Waldenstrom macroglobulinemia** (9761/3) when there is

- Clinical diagnosis of Waldenstrom macroglobulinemia **AND/OR**
- IgM monoclonal gammopathy in the blood **and/or** bone marrow

Note: There may be a mention of lymphoplasmacytic lymphoma (LPL) in the bone marrow biopsy or blood. LPL is an NOS code and Waldenstrom Macroglobulinemia is one of the two specific LPLs (Gamma heavy chain disease is the other).

Rule PH17 Code the primary site to the **involved bone marrow, lymph nodes, or tissue** and the histology **lymphoplasmacytic lymphoma** (9671/3) when

- There is a clinical diagnosis of lymphoplasmacytic lymphoma **AND/OR**
- Flow cytometry on bone marrow, lymph node(s),or tissue is positive for IgG, IgA and IgM monoclonal gammopathy

Module 7: Coding Primary Site for: Lymphomas, Extraosseous Plasmacytomas, Mast cell sarcoma, Histiocytic and Dendritic cell neoplasms, Heavy Chain Disease, Myeloid Sarcoma and Post-transplant lymphoproliferative disease (PTLD) PH18 – PH27

9590/3-9729/3, 9734/3, 9735/3-9738/3, 9740/3, 9751/3, 9755/3-9759/3, 9762/3, 9811/3-9818/3, 9823/3, 9827/3, 9837/3, 9930/3, 9971/3

Note 1: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3

Note 2: Do not simply code the site of a biopsy; use the information available from scans to determine the correct primary site

Rule PH18 Code the primary site to the specified **lymph node region** when the **site of lymphoma is described only as a mass**

- Mediastinal lymph nodes (C771) when the site of the lymphoma is described only as a **mediastinal mass**
- Intra-abdominal lymph nodes (C772) when the site of the lymphoma is described only as a **retroperitoneal mass or mesenteric mass**
- Inguinal lymph nodes (C774) when the site of the lymphoma is described only as an **inguinal mass**
- Pelvic lymph nodes (C775) when the site of the lymphoma is described only as a **pelvic mass**

Rule PH19 Code the primary site to the **specific lymph node region** when only **one lymph node or one lymph node region** is involved.

Rule PH20 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.

Note 1: Use this rule when there is bilateral involvement of lymph nodes.

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

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

Example 2: Code involvement of cervical lymph node chain and mandibular lymph node chain to lymph nodes of head, face and neck (C770).

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

Rule PH21 Code the **primary site** to **multiple lymph node regions, NOS (C778)** when multiple lymph node regions, 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: See Rule PH24 when there is also **organ** involvement.

Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Primary Site Coding Instructions for more information on coding primary site for lymphoma.

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

Example 1: Cervical (C770) and intra-thoracic (C771) lymph nodes involved with B-cell lymphoma. No indication of site of origin. Code the primary site to lymph nodes of multiple regions (C778).

Example 2: Biopsy of an axillary lymph node (C773) confirmed lymphoma. CT scans showed involvement of the axillary lymph nodes (C773) and the pelvic lymph nodes (C775). No additional involvement was identified during the work-up and no indication of site of origin. Code the primary site to lymph nodes of multiple regions (C778).

Rule PH22 Code the **primary site** to **lymph nodes, NOS** (C779) when:

- Lymphoma is present in an organ and lymph nodes that are **not** regional for that organ and the origin of the lymphoma cannot be determined even after consulting the physician **OR**
- Lymphoma is present in more than one organ and the regional nodes for all organs involved **OR**
- More than one organ and some combination of regional and distant nodes for the organs involved **OR**
- Lymph node(s) and organ(s) involved and no primary site/particular lymph node region is identified
- Lymph node(s) are involved and no primary site/particular lymph node region is identified

Note 1: Lymphoma can spread from organs to regional lymph nodes, but does not spread from the organ directly to distant lymph nodes.
Note 2: Use the Collaborative Stage Data Collection System to determine regional vs. distant lymph nodes.
Note 3: See Appendix C for help identifying lymph node names, chains, regions, and codes.
Note 4: Use this rule when there is no available information concerning where the lymphoma originated, such as historical cases.
Example 1: The patient has positive mediastinal lymph nodes (C771) and cervical lymph nodes (C770) and involvement of the stomach (C169). No further information is available. Code to lymph node, NOS (C779).
Example 2: Lymphoma is found in both lymph nodes and bone marrow. The pathology report is not available to help determine the primary site and no further information can be obtained. Code to lymph nodes, NOS (C779).
Example 3: The patient has a paraspinal soft tissue mass positive for lymphoma and no other areas of involvement. If the site of origin cannot be determined, code to lymph nodes, NOS (C779).
Example 4: The patient has involvement of two extranodal sites and regional lymph nodes for only one of those sites. If the site of origin cannot be determined, code the primary site to lymph nodes, NOS (C779).
Example 5: The patient has a history of Stage II lymphoma. No other information is available. Code to lymph nodes, NOS (C779).

Rule PH23 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 an organ. In rare cases a lymphoma may spread from lymph nodes to an extranodal site or extralymphatic organ by direct extension.

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 PH24 Code the **primary site** to the **organ** when lymphoma is present only in an **organ.**

Note 1: Includes lymphomas that are primary in the spleen. Splenic primaries are rare. Histologies that arise in the spleen include splenic marginal zone lymphoma (9689/3); hepatosplenic T-cell lymphoma (9716/3); splenic B-cell lymphoma/leukemia, unclassifiable (9591/3); splenic diffuse red pulp small B-cell lymphoma (9591/3); splenic marginal zone diffuse variant (9591/3); splenic EBV-associated B-cell lymphoproliferative disorder (9680/3). Follow-back for additional information when the histology is other than those listed **AND**

- The ***only information is a biopsy of the spleen OR***
- There is a physician statement that the spleen is the organ of origin

Note 2: Secondary involvement of distant lymph nodes and/or bone marrow are coded in CS extension.
Example: Pathology from stomach resection shows lymphoma. No other sites of involvement are identified. Code the primary site to stomach, NOS (C169).

Rule PH25 Code the primary site to the organ when a lymphoma is present in an **organ** and that **organ's regional lymph nodes**.

Note 1: In Stage II, III and IV disease, distant lymph nodes or other organs, such as spleen, may be involved. Disregard the distant lymph nodes and splenic involvement.
Note 2: Code the primary site to the organ. Use the Collaborative Stage Data Collections System to determine regional vs. distant lymph nodes.
Note 3: Secondary involvement of distant lymph nodes and/or bone marrow are coded in CS extension.
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 to spleen (C422).

Rule PH26 Code the **primary site** to **bone marrow** (C421) when lymphoma is **present only in the bone marrow** and/or **peripheral blood.**

Note 1: All available physical exams, scans, and other work-up must be negative for lymph node, tissue, or organ involvement **OR** no other workup was done.
Note 2: Code primary site to C421 when the only information available is a positive peripheral blood smear.
Example: Bone marrow biopsy is positive for diffuse B-cell lymphoma (DLBCL). No other work up performed. Code primary site to bone marrow. If further workup is done that identifies a primary site, reassign primary site.

Rule PH27 Code **primary site** to **unknown** primary site (C809) 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) OR multiple organ involvement without any nodal involvement.** See ICD-O-3 Rule D.

Module 8: NOS and More Specific Histology PH28 – PH29

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

Rule PH28 Code the **non-specific** (**NOS**) histology when the diagnosis is:

- **One non-**specific histology **AND**
- **Two or more specific** histologies **AND**
- The Heme DB Multiple Primaries Calculator documents the specific histologies and NOS are the **same primary AND**
- There is no further information regarding the physician's final diagnosis

Note 1: Use the Heme DB Multiple Primaries Calculator to confirm that the NOS and specific histologies are the same primary.
Note 2: See Instructions #3-5 in the Histology Coding Rules for cases with ambiguous terminology.
Example 1: The diagnosis is myelodysplastic/myeloproliferative neoplasm unclassifiable (9975/3), polycythemia vera (9950/3), essential thrombocythemia (9962/3). The Heme DB Multiple Primaries Calculator shows that myelodysplastic/myeloproliferative neoplasm unclassifiable and polycythemia vera are the same primary. The Multiple Primaries Calculator also shows that myelodysplastic/myeloproliferative neoplasm unclassifiable and essential thrombocythemia are the same primary. Follow-back produces no additional information. Code the histology myelodysplastic/myeloproliferative disorder, NOS (9975/3).
Example 2: Pathology report states morphologic features and immunophenotype of low grade B-cell lymphoma are most compatible with lymphoplasmacytic lymphoma or marginal zone lymphoma. The term "compatible with LPL (9671/3) or MZL (9699/3)" means that the immunophenotype was not definitely diagnostic for either specific disease. Default to the NOS diagnosis, the B-cell lymphoma, (9591/3).

Rule PH29 Code the **specific** histology when the diagnosis is:

- **One non**-specific (NOS) histology **AND**
- **One specific** histology **AND**
- The Heme DB Multiple Primaries Calculator documents the specific histology and NOS are the **same primary**

Note 1: See Instructions #3-5 in the Histology Coding Rules for cases with ambiguous terminology. If the specific histology diagnosis includes ambiguous terminology, code the NOS histology.

Note 2: Use the Heme DB Multiple Primaries Calculator to confirm that the NOS and specific histology are the same primary.

Module 9: Coding Primary Site and Histology PH30 – PH31

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

Use Only When Modules 1-8 are Not Applicable

Rule PH30 Use the Heme DB to determine the primary site and histology when rules PH1-PH29 do **not** apply.

Note: For primary site, use the information under Primary Site(s) in the Heme DB **and/or** the Abstractor Notes as instructed in the introduction of this manual.

Rule PH31 Code the histology to **the numerically higher** ICD-O-3 code when the histology code cannot be determined using the Heme DB.

Note: This rule should **rarely be used**.

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

Grade of Tumor Rules

Instructions for coding Grade/Differentiation were revised for **solid** tumor cases diagnosed 1/1/2014 and later. There were no changes to the Hematopoietic Grade rules below.

Note 1: A grade coding instruction is provided for each histology in the Heme DB based on the Grade of Tumor Rules below. The rules in the manual are the primary source for the grade rules. When applicable, the Heme DB can be used for a quick reference. Use of either the Heme DB grade coding instruction or the Grade of Tumor Rule will result in the same grade code.

Note 2: The only valid grade codes for hematopoietic neoplasms are 5 (T-cell), 6 (B-cell), 7 (Null cell), 8 (NK cell), and 9 (unknown).

Note 3: When there is no grade coding instruction, grade rule, or physician statement, code Grade/Phenotype "9" for unknown.

Note 4: Code the grade as indicated in the Heme DB and the Grade of Tumor Rules when the pathology report states a different grade than the one noted in the Heme DB or the Grade of Tumor Rules. **The grade instructions/rules take priority.**

Note 5: Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade for primaries diagnosed after 01/01/2010. This table is outdated. ***Table 13 may be used for cases diagnosed prior to 2010.***

Note 6: For those histologies that do not have a default grade (5-9), use a physician's statement to code the phenotype in the grade field, use statements from **any part** of medical record including but not limited to

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

If no default grade or physician's statement for grade is documented, then assign "9" for unknown.

Note 7: 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. Do not code grade 1, 2 or 3 describing follicular lymphomas.

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: Solitary mastocytoma of skin
9741/3: Systemic mastocytosis
9742/3: Mast cell leukemia
9751/3: Langerhans cell histiocytosis
9755/3: Histiocytic
9756/3: Langerhans cell sarcoma
9757/3: Interdigitating dendritic cell sarcoma
9758/3: Follicular dendritic cell sarcoma
9759/3: Fibroblastic reticular cell tumor
9801/3: Acute undifferentiated leukemia
9805/3: Acute biphenotypic leukemia
9806/3: Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); *BCR-ABL1*
9807/3: Mixed phenotype acute leukemia with t(v;11q23); *MLL* rearranged
9808/3: Mixed phenotype acute leukemia, B/myeloid, NOS

(list continued on next page)

9809/3: Mixed phenotype acute leukemia, T/myeloid, NOS
9875/3: Chronic myelogenous leukemia, *BCR-ABL1* positive
9876/3:Atypical chronic myeloid leukemia, *BCR-ABL1* negative
9945/3: Chronic myelomonocytic leukemia
9946/3: Juvenile myelomonocytic leukemia
9950/3: Polycythemia vera
9961/3: Primary myelofibrosis
9962/3: Essential thrombocythemia
9963/3: Chronic neutrophilic leukemia
9964/3: Chronic eosinophilic leukemia, NOS
9975/3: Myelodysplastic/myeloproliferative neoplasm, unclassifiable
9980/3: Refractory anemia
9982/3: Refractory anemia with ring sideroblasts
9983/3: Refractory anemia with excess blasts
9985/3: Refractory cytopenia with multilineage dysplasia
9986/3: Myelodysplastic syndrome associated with isolated del(5q)
9989/3: Myelodysplastic syndrome, unclassifiable
9991/3: Refractor neutropenia
9992/3: Refractory thrombocytopenia

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 or the Heme DB.

Rule G2 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.**

9700/3: Mycosis Fungoides
9701/3: Sezary's disease
9702/3: Peripheral T-cell lymphoma, NOS
9705/3: Angioimmunoblastic T-cell lymphoma
9708/3: Subcutaneous panniculitis-like T-cell lymphoma
9709/3: Primary cutaneous T-cell lymphoma
9714/3: Anaplastic large cell lymphoma, *ALK-positive* (unless pathologist specifically designates as a B-cell [code 6]
9716/3: Hepatosplenic T-cell lymphoma
9717/3: Enteropathy-associated T-cell lymphoma
9718/3: Primary cutaneous anaplastic large cell lymphoma
9724/3: Systemic EBV-positive T-cell lymphoproliferative disease of childhood
9725/3: Hydroa vacciniforme-like lymphoma
9726/3: Primary cutaneous gamma-delta T-cell lymphoma
9827/3: Adult T-cell leukemia/lymphoma
9834/3: T-cell prolymphocytic leukemia
9837/3: T lymphoblastic leukemia/lymphoma

(notes continued on next page)

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 G3 Code B-cell, **code 6**, for the following **B-cell precursor lymphoid neoplasms and the mature B-cell neoplasms**

9591/3: Non-Hodgkin lymphoma, NOS
9596/3: B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma
9597/3: Primary cutaneous follicle centre lymphoma
9659/3: Nodular lymphocyte predominant Hodgkin lymphoma
9670/3: Malignant lymphoma, small B lymphocytes, NOS
9671/3: Lymphoplasmacytic lymphoma
9673/3: Mantle cell lymphoma
9678/3: Primary effusion lymphoma
9679/3: Primary mediastinal (thymic) large B-cell lymphoma
9680/3: Diffuse large B-cell lymphoma (DLBCL)
9684/3: Malignant lymphoma, large B-cell, diffuse, immunoblastic, NOS
9687/3: Burkitt lymphoma
9688/3: T-cell/histiocyte-rich large B-cell lymphoma
9689/3: Splenic marginal zone lymphoma
9690/3: Follicular lymphoma
9691/3: Follicular lymphoma, grade 2
9695/3: Follicular lymphoma, grade 1
9698/3: Follicular lymphoma, grade 3
9699/3: Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)
9712/3: Intravascular large B-cell lymphoma
9728/3: Precursor B-cell lymphoblastic lymphoma
9731/3: Solitary plasmacytoma of bone
9732/3: Plasma cell myeloma
9734/3: Extraosseous plasmacytoma
9737/3: ALK-positive large B-cell lymphoma
9738/3: Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease
9761/3: Waldenstrom macroglobulinemia
9762/3: Heavy chain disease
9811/3: B lymphoblastic leukemia/lymphoma, NOS
9812/3: B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); *BCR-ABL1*
9813/3: Lymphoblastic leukemia/lymphoma with t(v;11q23); *MLL*
(list continues on next page)

9814/3: B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); *TEL-AML1 (ETV6-RUNX1)*
9815/3: B lymphoblastic leukemia/lymphoma with hyperdiploidy
9816/3: B lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL
9817/3: B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); *IL3-IGH* (
9818/3: B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); *E2A-PBX1 (TCF3-PBX1)*
9823/3: Chronic lymphocytic leukemia/small lymphocytic lymphoma
9826/3: Burkitt cell leukemia
9833/3: B-cell prolymphocytic leukemia
9940/3: Hairy cell leukemia

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 (NK-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 Tables B7 and B8 in Appendix B or the Heme DB.

Rule G4 Code NK-cell (natural killer cell), code 8, for the following neoplasms:

9719/3: Extranodal NK-/T-cell lymphoma, nasal type
9948/3: Aggressive NK-cell leukemia

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 or the Heme DB.

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 and T-cell.**

Rule G6 Code B-cell, **code 6**, when the neoplasm is identified as **B-cell, B-cell phenotype, B-precursor, pre-B, or null-cell and B-cell.**

Rule G7 Code Null cell, non-T non-B, **code 7**, when the neoplasm is described as **null cell, non-T non-B,** or **common cell.**

Rule G8 Code Natural Killer (NK) cell, **code 8**, when the neoplasm is described as **NK cell, natural killer cell, nasal NK/T-cell lymphoma, or null-cell and NK cell.**

Rule G9 Code cell type not determined, not stated, not applicable, **code 9**, when Rules G1 – G8 do not fit the case **AND**

- 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**

Appendix A
History of Hematopoietic and Lymphoid Neoplasm Coding

History of Coding Lymphoid Tissue and Hematopoietic System Neoplasms

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 characteristics 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 B.

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 are based on ICD-O-3 term and category assignment only. Obsolete designations have not been updated in ICD-O-3 to match the 2008 WHO Classification. Revised obsolete 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, lymphocytic predominance, diffuse	*Source: Lukes-Butler classification, 1966; Obsolete: REAL classification 1994*	9651/3	Lymphocyte-rich classical Hodgkin lymphoma
Hodgkin disease, lymphocytic predominance, NOS	*Source : Rye classification, 1966 ; Obsolete : REAL classification 1994*	9651/3	Lymphocyte-rich classical Hodgkin lymphoma
Hodgkin disease, lymphocytic-histiocytic predominance	*Source: Lukes-Butler classification, 1966; Obsolete as of REAL classification 1994*	9651/3	Lymphocyte-rich classical Hodgkin lymphoma
Hodgkin paragranuloma	*Source : Jackson-Parker Classification, 1944 ; Obsolete : 1966*	9659/3	Nodular lymphocyte predominant Hodgkin lymphoma
Hodgkin paragranuloma, NOS	*Source : Jackson-Parker Classification, 1944 ; Obsolete : 1966*	9659/3	Nodular lymphocyte predominant 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		

(Table continued on next page)

Table A3: Lymphoid Neoplasm Obsolete Terms (continued)

Classification	Obsolete Term	Current Term	ICD-O-3 Code
T-Cell and NK-Cell Neoplasms	Angiocentric T-cell lymphoma	Extranodal NK/T cell lymphoma, nasal type	9719/3
	Malignant reticulosis, NOS		
	Malignant midline reticulosis		
	Polymorphic reticulosis		
	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 erythremia in ICD-O-1 9840/3*	Acute erythroid leukemia	9840/3
	Acute erythremic myelosis	*Listed as synonym for acute erythremia 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 erythremia	*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, unclassifiable	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

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
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
Cutaneous mastocytosis (solitary mastocytoma of skin)	9740/1
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 neoplasms with *FGFR1* abnormalities	9967/3
Myeloid and lymphoid neoplasms with *PDGFRA* rearrangement	9965/3
Myeloid neoplasms 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 with multilineage dysplasia	9985/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3
Childhood myelodysplastic syndrome Refractory cytopenia of childhood	9985/3

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(3)(q21;q26.2) or t(3;3)(q21;q26.2); *RPN1-EVI1*	9869/3
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); *CBFB-MYH11*	9871/3
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 promyelocytic leukemia with t(15;17)(q22;q12), *PML-RARA*	9866/3
Acute myeloid leukemia with myelodysplasia-related changes	9895/3
Therapy-related myeloid neoplasm	9920/3
Acute myeloid leukemia, NOS	9861/3
Acute basophilic leukemia	9870/3
Acute erythroid leukemia	9840/3
Acute megakaryoblastic leukemia	9910/3
Acute monoblastic and monocytic leukemia	9891/3
Acute myeloid leukemia with maturation	9874/3
Acute myeloid leukemia with minimal differentiation	9872/3
Acute myeloid leukemia without maturation	9873/3
Acute myelomonocytic leukemia	9867/3
Acute panmyelosis with myelofibrosis	9931/3
Myeloid sarcoma	9930/3

WHO Preferred Term	ICD-O-3
Myeloid proliferations related to Down syndrome	*No Code*
Myeloid leukemia associated with Down syndrome	9898/3
Transient abnormal myelopoiesis	9898/1
Blastic plasmacytoid dendritic cell neoplasm	9727/3

Table B6: Acute Leukemias of Ambiguous Lineage

WHO Preferred Term	ICD-O-3
Acute undifferentiated leukemia	9801/3
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
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

Note: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3.

WHO Preferred Term	ICD-O-3
T lymphoblastic leukemia/lymphoma	9837/3
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(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(12;21)(p13;q22); *TEL-AML1 (ETV6-RUNX1)*	9814/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); *MLL* rearranged	9813/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3

Table B8: Mature B-Cell Neoplasms

Note: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3.

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 - B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma - DLBCL associated with chronic inflammation - EBV positive DLBCL of the elderly - Primary cutaneous DLBCL, leg type - Primary DLBCL of the CNS	9680/3
Extranodal marginal zone lymphoma of mucosal-associated lymphoid tissue (MALT lymphoma) - Nodal marginal zone lymphoma - Pediatric nodal marginal zone lymphoma	9699/3
Extraosseous plasmacytoma	9734/3
Follicular lymphoma - Pediatric follicular lymphoma	9690/3
Follicular lymphoma, grade 1	9695/3
Follicular lymphoma, grade 2	9691/3
Follicular lymphoma, grade 3	9698/3
Hairy cell leukemia	9940/3
Heavy chain diseases - Alpha heavy chain disease - Gamma heavy chain disease - Mu 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

(Table continued on next page)

Table B8: Mature B-Cell Neoplasms (continued)

WHO Preferred Term	ICD-O-3
Splenic B-cell lymphoma/leukemia, unclassifiable - Hairy cell leukemia-variant - Splenic disuse red pulp small B-cell lymphoma	9591/3
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 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

Note: For the purpose of using the rules, a non-Hodgkin lymphoma is any lymphoma (including the leukemia/lymphomas) not stated to be Hodgkin lymphoma, NOS or a type of Hodgkin lymphoma.

- Hodgkin lymphomas are: 9650/-9653/3, 9655/3, 9659/3, 9663/3.

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
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 - Anaplastic large cell lymphoma, *ALK negative*	9702/3
Primary cutaneous anaplastic large cell lymphoma	9718/3
- Primary cutaneous CD4 positive small/medium T-cell lymphoma - Primary cutaneous CD8 positive aggressive epidermitropic cytotoxic 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

(Table continued on next page)

Table B9: Mature T-Cell and NK-Cell Neoplasms (continued)

WHO Preferred Term	ICD-O-3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
T-cell large granular lymphocytic leukemia - Chronic lymphoproliferative disorder of NK-cells	9831/3
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 xanthogranuloma	No Code
Fibroblastic reticular cell tumor	9759/3
Follicular dendritic cell sarcoma	9758/3
Histiocytic sarcoma	9755/3
Interdigitating dendritic cell tumor - 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
Classical Hodgkin lymphoma type PTLD	*
Early lesions	*No Code*
Plasmacytic hyperplasia	9971/1
Infectious mononucleosis-like PTLD	9971/1
Polymorphic PTLD	9971/3
Monomorphic PTLD (B- and T/NK-cell types)	*

*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
***The right and left are separate regions per AJCC**

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
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	C772	Intra-abdominal	Para-aortic
Aortico-pulmonary window (subaortic)	C772	Intra-abdominal	Para-aortic
Appendiceal	C772	Intra-abdominal	Mesenteric
Apical axillary; deep axillary (Level III axillary)	C773	Axilla or arm	Axillary, right and left*
Ascending aortic lateral	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, anterior	C773	Axilla or arm	Axillary, right and left*
Axillary, lateral	C773	Axilla or arm	Axillary, right and left*
Axillary, Level I (low axillary, superficial axillary)	C773	Axilla or arm	Infraclavicular, right and left*
Axillary, Level II	C773	Axilla or arm	Infraclavicular, right and left*
Axillary, Level III (apical axillary, deep axillary)	C773	Axilla or arm	Infraclavicular, right and left*
Azygos (lower paratracheal)	C771	Intrathoracic	Mediastinal
Brachial	C773	Axilla or arm	Axillary, right and left*
Brachiocephalic	C773	Axilla or arm	Axillary, right and left*
Bronchial	C771	Intrathoracic	Hilar
Bronchopulmonary	C771	Intrathoracic	Hilar

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Buccal	C770	Head, face and neck	Cervical, right and left*
Buccinator (facial)	C770	Head, face and neck	Cervical, right and left*
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 axillary (Level III axillary)	C773	Axilla or arm	Infraclavicular, right and left*
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; anterior to thyroid isthmus	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

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
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
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, right and left*
Hilar; bronchopulmonary, proximal lobar, pulmonary root	C771	Intrathoracic	Hilar, right and left*
Hilar, hepatoduodenal ligament	C772	Intra-abdominal	Para-aortic
Hilar, pulmonary root	C771	Intrathoracic	Hilar, right and left*
Hypogastric	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 hemorrhoidal	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
Infundibulopelvic (Utero-ovarian)	C775	Pelvic	Pelvic, right and left*
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*
Interaortocaval	C772	Intra-abdominal	Para-aortic

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Intercostal	C771	Intrathoracic	Mediastinal
Interlobar (within the lung)/intrapulmonary	C771	Intrathoracic	Mediastinal
Internal iliac	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
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)	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
Low axillary (Level I axillary)	C773	Axilla or arm	Infraclavicular, right and left*
Lower jugular	C770	Head, face and neck	Cervical, right and left*
Lower paratracheal	C771	Intrathoracic	Mediastinal
Lower periesophageal (intrathoracic esophagus)	C771	Intrathoracic	Mediastinal
Lower peritracheal	C771	Intrathoracic	Mediastinal

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
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
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*

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Parametrial	C775	Pelvic	Pelvic, right and left*
Parapancreatic	C772	Intra-abdominal	Para-aortic
Parapharyngeal	C770	Head, face and neck	Cervical, right and left*
Pararectal (anorectal)	C775	Pelvic	Pelvic, 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
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*

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Posterior cecal	C772	Intra-abdominal	Para-aortic
Posterior cervical	C770	Head, face and neck	Cervical, right and left*
Posterior mediastinal	C771	Intrathoracic	Mediastinal
Posterior triangle	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*
Presymphseal	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
Rectal	C775	Pelvic	Pelvic, right and left*
Recurrent laryngeal	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*

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
Rosenmuller's-Cloquet's nodes	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
Sister Mary Joseph	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 (lineal)	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*
Suboccipital	C770	Head, face and neck	Cervical, right and left*
Subpleural (in the periphery of the lung)	C771	Intrathoracic	Mediastinal
Subpyloric (infrapyloric)	C772	Intra-abdominal	Para-aortic
Subscapular (posterior axillary)	C773	Axilla or arm	Axillary, right and left*
Substernal	C771	Intrathoracic	Mediastinal
Superficial axillary (Level I axillary)	C773	Axilla or arm	Infraclavicular, right and left*
Superficial inguinal	C774	Inguinal region or leg	Inguino-femoral, right and left*
Superior gastrocolic (left gastrocolic)	C772	Intra-abdominal	Mesenteric
Superior hemorrhoidal	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*

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O-3 Lymph Node Region(s)	AJCC/CS Staging
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	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 deep cervical (internal jugular)	C770	Head, face, and neck	Cervical, right and left*
Upper jugular	C770	Head, face and neck	Cervical, right and left*
Utero-ovarian (infundibulopelvic)	C775	Pelvic	Pelvic, right and left*
Virchow's node (left 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 are effective 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); *RPN1-EVI1*	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(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(12;21)(p13;q22); *TEL-AML1 (ETV6-RUNX1)*	9814/3
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 *Note: Alternate name for 9680/3 in ICD-O-3*	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 neoplasms with *FGFR1* abnormalities	9967/3
Myeloid and lymphoid neoplasms with *PDGFRA* rearrangement	9965/3
Myeloid neoplasms with *PDGFRB* rearrangement	9966/3
Myeloid leukemia associated with Down Syndrome	9898/3
Plasmablastic lymphoma *Note: Alternate name for 9684/3 in ICD-O-3*	9735/3

New Histology Term	ICD-O Code
Polymorphic PTLD	9971/3
Primary cutaneous follicle centre lymphoma	9597/3
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-cell/histiocyte rich large B-cell lymphoma *Note: Alternate name for 9680/3 in ICD-O-3*	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 2010+ diagnosis. Column 1 is the new WHO code and column 2 is the new description for that code. 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 are effective 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.

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

ICD-O Code	New Histology Term
9818/3	B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); *E2A PBX1 (TCF3 PBX1)*
9865/3	Acute myeloid leukemia with t(6;9)(p23;q34); *DEK-NUP214*
9869/3	Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); *RPN1EVI1*
9898/3	Myeloid leukemia associated with Down Syndrome
9911/3	Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); *RBM15-MKL1*
9965/3	Myeloid and lymphoid neoplasms with *PDGFRA* rearrangement
9966/3	Myeloid neoplasms with *PDGFRB* rearrangement
9967/3	Myeloid and lymphoid neoplasms with *FGFR1* abnormalities
9971/3	Polymorphic PTLD
9991/3	Refractory neutropenia
9992/3	Refractory thrombocytopenia

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 *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues*, 4th Edition published in 2008. Reporting of these neoplasms is effective with cases diagnosed 1/1/2010 and after. There are no plans or mandates to collect 2008 and 2009 cases having these diagnoses.

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

Appendix E
Obsolete Hematopoietic Neoplasm Codes

The table below is based on the 2008 WHO (see Obsolete Hematopoietic Histology Codes). The table lists the obsolete ICD-O-3 codes, the obsolete descriptions, the current ICD-O-3 codes (per the Hematopoietic database), and the current descriptions. All these codes were obsolete effective 12/31/2009. The current ICD-O-3 code should be used starting with 1/1/2010 cases.

Note: Effective January 2015, any cases with an obsolete ICD-O-3 code will be converted to the current ICD-O-3 code. None of the obsolete codes will be used for 2010 and forward.

Obsolete ICD-O-3 Code	Obsolete Description	Current ICD-O-3 Code	Current Description
9654/3	Hodgkin lymphoma, lymphocyte depletion, diffuse fibrosis	9653/3	Lymphocyte-depleted classical Hodgkin lymphoma
9661/3	Hodgkin granuloma	9650/3	Classical Hodgkin lymphoma
9662/3	Hodgkin sarcoma	9650/3	Classical Hodgkin lymphoma
9664/3	Hodgkin lymphoma, nodular sclerosis, cellular phase	9663/3	Nodular sclerosis classical Hodgkin lymphoma
9665/3	Hodgkin lymphoma, nodular sclerosis, grade 1	9663/3	Nodular sclerosis classical Hodgkin lymphoma
9667/3	Hodgkin lymphoma, nodular sclerosis, grade 2	9663/3	Nodular sclerosis classical Hodgkin lymphoma
9670/3	Malignant lymphoma, small B lymphocytic, NOS	9823/3	CLL/small lymphocytic lymphoma
9675/3	Malignant lymphoma, mixed small and large cell, diffuse	9690/3	Follicular lymphoma
9684/3	Malignant lymphoma, large B-cell, diffuse, immunoblastic, NOS	9680/3	Diffuse large B-cell lymphoma (DLBCL), NOS
9728/3	Precursor B-cell lymphoblastic lymphoma	9811/3	B lymphoblastic leukemia/lymphoma, NOS
9729/3	Precursor T-cell lymphoblastic lymphoma	9837/3	T lymphoblastic leukemia/lymphoma
9733/3	Plasma cell leukemia	9732/3	Plasma cell myeloma
9750/3	Malignant histiocytosis	9751/3	Langerhans cell histiocytosis
9752/1	Langerhans cell histiocytosis, unifocal	9751/3	Langerhans cell histiocytosis
9753/1	Langerhans cell histiocytosis, multifocal	9751/3	Langerhans cell histiocytosis
9754/3	Langerhans cell histiocytosis, disseminated	9751/3	Langerhans cell histiocytosis
9760/3	Immunoproliferative disease, NOS	-	See codes 9761/3 and 9762/3
9764/3	Immunoproliferative small intestinal disease	9762/3	Heavy chain disease
9805/3	Acute biphenotypic leukemia	-	Assign to one of the new codes in the 9806-9809 range. For acute biphenotypic leukemia, NOS, assign code: 9809/3
9835/3	Precursor cell lymphoblastic leukemia, NOS	9811/3	B lymphoblastic leukemia/lymphoma, NOS
9836/3	Precursor B-cell lymphoblastic leukemia	9811/3	B lymphoblastic leukemia/lymphoma, NOS
9960/3	Chronic myeloproliferative disease, NOS	9975/3	Myelodysplastic/myeloproliferative neoplasm unclassifiable
9984/3	Refractory anemia with excess blasts in transformation	9983/3	Refractory anemia with excess blasts
9987/3	Therapy related myelodysplastic syndrome, NOS	9920/3	Therapy-related myeloid neoplasms