Navigating Heme-Lymph database

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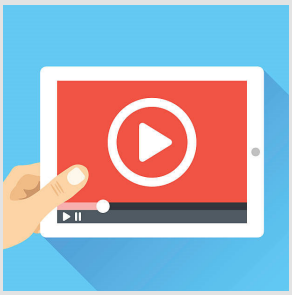
SHRI Video Training Series | Iowa Cancer Registry

Recorded 2/2023



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Change Log

- Revision History for the Hematopoietic Project - SEER Registrars (cancer.gov)

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- **CHOOSE THE RIGHT YEAR**
  - **New histologies**
  - **Obsolete histologies**
  - **Changes in behavior**
- **In H-L database:**

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Steps for using Heme DB

- Update to “Steps in Priority Order for Using the Heme DB and Hematopoietic Coding Manual”
- Search function has recently changed

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Step 1: Identify working histology code

a. Search the Heme DB

b. “Show Alternate Names”: This box appears under the Search box. If this box is checked, the results will include an additional column that shows where alternate names include the words being search

c. Search on histology code if desired, i.e., 9867/3.

d. When multiple results are displayed, click on the desired term (e.g. acute myelomonocytic leukemia) to display the record.

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Step 2: Use MPH Rules

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

b. Use the Hematopoietic Multiple Primaries Calculator in the Heme DB **only when instructed** by the rules in the Hematopoietic Manual.

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Step 3: Use PH Rules to verify or revise histology

a. When the PH rules lead you to a different histology code, enter that code in the Heme DB search box and display the record for that histology

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

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Step 4: Determine primary site

a. See Primary Site Coding Instructions.

b. For certain histologies, only one primary site code is displayed in the Heme DB

- i. The primary site code displayed under Primary Site(s) is the only site code to be used for that histology

c. When there is no primary site code listed under Primary Site(s) in the Heme DB

- 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.

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Step 5: Determine Grade

- Note: Grade is no longer collected for cases diagnosed 1/1/2018 and forward
- 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

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Primary Site

- Per Jennifer Ruhl: Not all sites of involvement are used to assign primary sites. Some sites of disease may be metastatic.
- Module 7 (PH18-PH27)
- Note 2: Do not simply code the site of a biopsy; use the information available from imaging to determine the correct primary site
- Note 3: Secondary involvement of distant lymph nodes (for an extranodal lymphoma), bone marrow, liver, spleen or CNS are included in the stage fields only. This secondary involvement excludes rare primary lymphoid neoplasms of spleen, multifocal lung involvement, liver or CNS (see PH Rules). **Secondary involvement of distant site(s) is disregarded for the purpose of coding primary site.** For lymphoid neoplasms, this secondary or distant involvement is akin to metastasis for solid tumors and does not alter the primary site assigned by the physician or determined using the PH Rules.

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Case Reportability Instructions

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Reportability

1. Search the Heme DB to determine case reportability.
2. Report all cases with morphology code 9590-9993 with /3 behavior.
3. Report heme and lymph neoplasm with morphology 9590-9993 listed as /1 that are described as malignant by physician. **Note: Do not report in situ (/2) lymphomas.**
4. **Report the case when dx is preceded by ambig terms.**

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Ambiguous Terms for reportability

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

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Multiple Primary Rules

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General Instructions for Multiple Primary Rules

1. Start with M1 for each case, move through rules and stop at the first rule that applies. Use the M rule references in Heme DB as a guide only.
2. Within these rules, the term “chronic neoplasm” means that a neoplasm has the potential to transform into another, more acute neoplasm.
3. Common to have provisional diagnosis or several provisional (differential) NOS diagnoses that lead to more testing and a more specific dx. **These are not multiple primaries, just steps in the diagnostic workup.**
4. The Heme DB Multiple Primaries Calculator is to be used **only** when instructed to do so.

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MP Rules

M1 (S)	Single primary when minimal info available
M2 (S)	Single primary when there is a single histology *Exceptions, Notes, Examples*
M3 (S)	Single primary when sarcoma dx simultaneously with or after leukemia of same lineage
M4 (S)	Single primary when 2 or more types of NHL simultaneous in same anatomic location (same node, same organ, same tissue)
M5 (S)	Single primary when both HL and NHL simultaneous in same anatomic location (same node, same organ, same tissue)

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MP Rules

M6 (M)	Multiple primaries when HL in one location and NHL in another location.
M7 (S)	Single primary when more specific histology is dx after an NOS when Heme DB **MPC confirms the NOS and more specific are SAME primary.

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MP Rules

	[Transformation rules M8-M13]
M8 (S)	Single primary and code acute neoplasm when both chronic and acute dx simultaneously or within 21 days and only one biopsy (bm, LN, tissue).
M9 (S)	Single primary and code later dx when both chronic and acute are dx simultaneous or within 21 days and no available documentation on biopsy.
M10 (M)	Multiple primary when neoplasm originally dx as chronic neoplasm and second dx of acute >21 days after chronic.
M11 (M)	Multiple primary when both chronic and acute neoplasms are diagnosed simultaneously or within 21 days AND documentation of two biopsies: one confirms chronic and one confirms acute.

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MP Rules

	[Transformation rules M12-M13] Treatment related
M12 (S)	Single primary when neoplasm originally dx as acute and reverts to a related chronic neoplasm more than 21 days after acute and no confirmation available that patient was treated for acute. [Acute ➡ Chronic without treatment]
M13 (M)	Multiple primary when neoplasm originally dx as acute and reverts to a chronic after treatment. [Acute ➡ Chronic AFTER treatment]

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MP Rules

M14 (S)	Single primary when PTLD dx simultaneously with any B-cell lymphoma, T-cell lymphoma, HL or myeloma. Note 1: As of 2021,(PTLD) WITHOUT an associated lymphoma or plasmacytoma is a /1 and is no longer reportable Note 2: 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 as a second primary. • This note only applies when the PTLD is diagnosed 2010-2020. If the PTLD is diagnosed after 12/31/2020 and is followed by a lymphoma, the lymphoma will be a first primary (if no other cancers present)
M15	Use Heme DB **MPC to determine number of primaries for all cases that do not meet criteria of M1-M14.

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Primary Site Coding

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Instructions

1. Use these instructions, PH Rules and Heme DB to code primary site.
2. Do not use these primary sites for heme neoplasms:
   a) C423 Reticuloendothelial system, NOS
   b) C424 Hematopoietic system, NOS
3. Heme DB gives primary site:
   a) Use the specific site code listed, when applicable
   b) Primary site text field provides additional info
4. Code primary site using:
   a) Scans
   b) Medical record documentation
   c) Path report
   d) Heme DB

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Instructions

5. Secondary involvement of distant LNs (extranodal lymphoma,) bone marrow, liver, spleen or CNS are included in staging fields only. Disregard secondary involvement for purpose of coding primary site.

6. Code primary site as indicated on pages 36-40 for each category of disease.

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Histology Coding

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Histology Instructions

1. Code histology identified by Definitive Diagnostic Method(s) (may be any of following) and no hierarchy to this list:
   a) Clinical dx
   b) Genetic test
   c) Immunophenotyping
   d) Cytology
   e) Pathology (final dx, comment on final dx, addendum, CAP protocol/synoptic report)
2. When tests or reports defined as DDM are NOT AVAILABLE use this hierarchy:
   a) Documentation in med rec referring to original scans, genetic testing, immunophenotyping, or path report
   b) Documentation in med rec that refers to histology

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Instructions for coding Histology

3. When test or report lists a specific histology with ambiguous term and an NOS histology, **code the NOS histology.**

Note 1: Ambig used for reportability

Note 2: Ambig terms may not be used when specific histology has not been confirmed. If no further info re more specific histology, assign NOS.

See notes and examples

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Instructions for coding Histology

4. If one histology preceded by ambig term, review abstractor notes in Heme DB, look for other info to confirm dx.

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 relevant immunophenotyping or genetics is present in abstractor notes and the only histology preceded by ambig term, code ambig term so case can be reported for incidence.

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Primary Site & Histology Rules

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PH Coding Rules

1. Rules in 9 modules. Each module covers group of related heme or lymph neoplasms. May find neoplasm in more than one module.
2. Modules are not hierarchical. Rules within each module are in order. Apply rules in each module in order. Stop at the first rule that applies.

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PH Modules

Module 1	PH1	Post-Transplant Lymphoproliferative Disorder (PTLD) for 2010-2020 only.
Module 2	PH 2-4	Plasmacytomas (9734/3) (9731/3)
Module 3	PH 5-6	CLL/SLL (9823/3)
Module 4	PH 7-8	Leukemia/Lymphoma (numerous histologies)
Module 5	PH 9-10	Myeloid neoplasms (numerous histologies) Mast Cell Neoplasms (9740/3) (9742/3) (9930/3)
Module 6	PH 11-17	NHL (numerous histologies)
Module 7	PH18-27	Hodgkin Lymphomas; NHL; Extraosseous plasmacytomas etc (numerous)
Module 8	PH28-29	NOS and more specific Histology All heme and lymph neoplasms 9590/3-9993/3
Module 9	PH30-31	All. Use only when Modules 1-8 are not applicable

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PH RULES

	Module 7: HL and NHL
PH18	Applicable for Hodgkin & non-Hodgkin Lymphomas only: Code the primary site to the specified lymph node region when the site of lymphoma is described only as a mass.
	Note 1: This rule does not apply to other descriptions of "mass." For example, a "mass" in the neck is likely describing cervical lymph node involvement and does not meet the criteria for this rule.
	• 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

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PH RULES

	Module 3: CLL/SLL
PH5	Code the primary site to bone marrow (C421) when the bone marrow is involved or when only peripheral blood is involved.
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 AND no bone marrow involvement or when it is unknown if bone marrow is involved

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Appendices A-D

Appendix	
A	History of Hematopoietic and Lymphoid Neoplasm Coding
B	WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues | Histology Lineages
C	Lymph Node/Lymph Node Chain Reference Table
D	Introduction to Genetic Nomenclature

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Appendix B

Table B6: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms

WHO Preferred Term	ICD-O
Acute myeloid leukemias with recurrent genetic abnormalities	
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13.3;q13.1); *RBM15-MKL1*	9911/3
Acute myeloid leukemia with *BCR-ABL1* (2021)+	9912/3*
Acute myeloid leukemia with biallelic mutation of *CEBPA* (2021+)	9878/3*
Acute myeloid leukemia with inv(3)(q21.3q26.2) or t(3;3)(q21.3;q26.2); *GATA2, MECOM*	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 mutated *NPM1* (2021+)	9877/3*
Acute myeloid leukemia with mutated *RUNX1* (2021+)	9879/3*

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Appendix D Intro to Genetic Nomenclature

Pg 81-84 in Heme-Lymph Manual document

Selected Types of Abnormalities/Mutations

Mutation Type	Abbreviation(s)	Description	Nomenclature Example(s)
Insertion *	ins	Addition of DNA into a gene.	ins(18;5)(q21.1;q31.2)
Deletion	del	Removal of DNA; may occur in one or more base pairs, entire gene(s), or chromosome arm (p or q).	del(5q); del(6q21)
Duplication *	dup	DNA abnormally copied one or more times.	dup(21); FLT3-ITD (Where ITD = internal tandem duplication)
Inversion	inv	Rearrangement within a single chromosome in which a chromosome segment undergoes breakage and rearrangement within itself.	inv(16); inv(3); inv(16)(p13.1;q22); inv(3)(q21;q26.2) (Sometimes described as a translocation between a single chromosome: t(16;16)(p13.1;q22))
Translocation	t(x;x) **	Rearrangement between two chromosomes in which a chromosome segment breaks off and attaches to a different chromosome.	t(9;22); t(8;21); t(9;22)(q34;q11.2); t(8;21)(q22;q22)
Trisomy	(XY, +x) **	An extra copy (three total copies) of the specified chromosome.	47(XY,+8); Trisomy 21; Gain of chromosome 9 (Sometimes these are referred to as just "Trisomy" or "Gain of" abnormalities without abbreviation or specific karyotype notation.)
Monosomy	(XY,-x) **	The presence of only one chromosome from the specified chromosome pair.	45(XY,-16); Monosomy 7; Loss of chromosome 5 (Sometimes these are referred to as just "Monosomy" or "Loss of" abnormalities without abbreviation or specific karyotype notation.)

* Uncommon as a sole genetic/molecular abnormality documented in heme/lymphoid neoplasms.

** Where lowercase "x" represents the chromosome number involved.

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- Search
  - Term or code from appendix B
  - Acronyms or Genetics
    - RAEB
    - (p23;q34.1)
- Alternate names in database
  - Not all parts of genetic string are necessary
  - 9897/3 Example

Alternate Names

Acute myeloid leukemia, MLL
Acute myeloid leukemia with 11q23 (MLL) abnormalities
Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL
Acute myeloid leukemia with t(9;11)(p22;q23) resulting in KMT2A-MLLT3

- Genetics Data or Immunophenotyping
  - Terms positive or expression mean the same thing per Dr. Nashelsky

Immunophenotyping

BCL2 expression and positive
BCL6 positive
CD5 negative
CD10 expression and positive
CD19 expression
CD20 positive

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Thank You
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