



National Cancer Institute

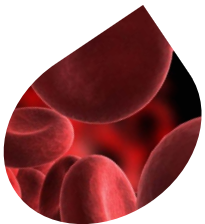
Hematopoietic and Lymphoid Neoplasm Project





Acknowledgments

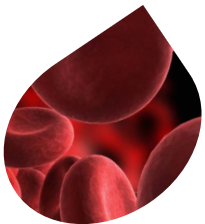
- American College of Surgeons (ACOS) Commission on Cancer (COC)
- Canadian Cancer Registries (CCR)
- National Cancer Registrars Association (NCRA)
- National Program of Cancer Registries (NPCR) of the Centers for Disease Control (CDC)
- North American Association of Central Cancer Registries (NAACCR)





With Special Thanks to

- Graca Dores, MD
- Charles Platz, MD
- Amy Blum, RHIT, CTR
- The Hematopoietic Working Group





Primary Site and Histology Rules

Part I

Carol Hahn Johnson, BS, CTR

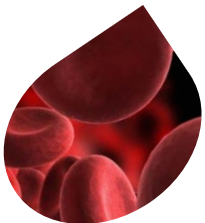
NCI SEER

September 2009



PH Rules

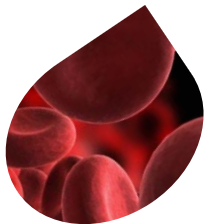
- Primary site and histology rules combined (PH)
- Rules apply to problematic
 - Primary sites
 - Histologies
 - Terms





Note 1

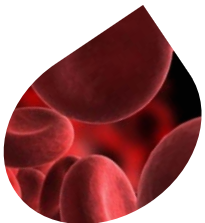
Use the Primary Site and Histology Rules
before using the Hematopoietic DB





Note 2

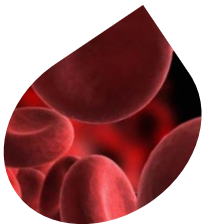
The primary site and histology coding rules are divided into nine modules. Each **module** covers a group of **related** hematopoietic or lymphoid **neoplasms**. However, a specific histology may be covered in more than one module





Note 3

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





Module Headers

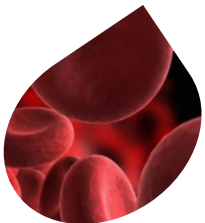
Module 2: Plasma Cell Neoplasms

Solitary plasmacytoma of bone 9731/3

**Plasma cell myeloma/multiple myeloma
9732/3**

Extraosseous plasmacytoma 9734/3

PH4-PH8





Index

Primary Site and Histology Rules Flowchart

Module 1: General Instructions 19

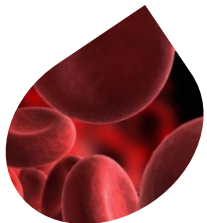
All Hematopoietic and lymphoid neoplasms 9959/3-9992/3

Module 2: Plasma Cell Neoplasms 21

Solitary plasmacytoma of bone 21

Plasma cell myeloma/multiple myeloma 21

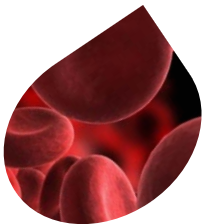
Extraosseous plasmacytoma





Note 4

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





Instructions Within Modules

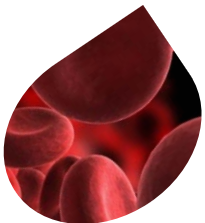
Module 1

Go to the appropriate Module 2-8.

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

Module 2

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





Module 1: General Instructions
All hematopoietic and lymphoid
neoplasms 9590/3-9992/3
PH1-PH3



Rule	Histology	Other	Code
PH1	All		Code primary site using <ul style="list-style-type: none">• Scans• Medical record documentation• Pathology report• Hematopoietic DB



Notes/Examples PH1

Notes / Examples

Note: For hematopoietic neoplasms the pathology report is not the automatic default standard for determining the primary site. The standard for determining primary site differs depending upon the specific histology.



Rule	Histology	Other	Code
PH2	All		<p>Code from definitive diagnostic method (See Hematopoietic DB). Definitive diagnostic method can be</p> <ul style="list-style-type: none">•Clinical diagnosis•Genetic test•Immunophenotyping•Cytology•Pathology<ul style="list-style-type: none">○ Final diagnosis○ Comment on final diagnosis○ Addenda to final diagnosis○ CAP protocol



Rule	Histology	Other	Code
PH3	When tests or reports defined as definitive diagnosis are not available		Code primary site and histology from medical practitioner's statement on <ul style="list-style-type: none">• Medical record• Death certificate



Notes / Examples

Go to the appropriate Module 2-8.

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



Module 2: Plasma Cell Neoplasms

Solitary plasmacytoma of bone

9731/3

**Plasma cell myeloma/multiple
myeloma 9732/3**

Extraosseous plasmacytoma

9734/3

PH 4—PH 8



Rule	Histology	Other	Code
PH4	Any of the following occur in a site other than bone : <ul style="list-style-type: none">• Plasmacytoma• Extraosseous or extramedullary plasmacytoma• Solitary plasmacytoma• Multiple plasmacytomas• Multiple extraosseous or extramedullary plasmacytomas		<ol style="list-style-type: none">1. Primary site to the site of origin (lymph node region(s), tissue, or organ)2. Histology extramedullary plasmacytoma (9734/3)

Notes / Examples

Note 1: Extramedullary and extraosseous mean not occurring in bone

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

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

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

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



Rule	Histology	Other	Code
PH5	Any of the following occur in bone <ul style="list-style-type: none">● Plasma cell neoplasm● Solitary plasmacytoma● Solitary plasmacytoma of bone● Solitary medullary plasmacytoma● Multiple plasmacytomas● Multiple plasmacytomas of bone● Multiple medullary plasmacytomas of bone● Multiple medullary plasmacytomas		<ol style="list-style-type: none">1. Primary site to the specific bone (C400-C419)2. Histology solitary plasmacytoma of bone (9731/3)



Notes / Examples

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

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



Rule	Histology	Other	Code
PH6		Only information is documentation that patient had a plasmacytoma or solitary plasmacytoma	<ol style="list-style-type: none">1. Primary site unknown (C809)2. Histology solitary plasmacytoma of bone (9731/3)



Notes / Examples

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



Rule	Histology	Other	Code
PH7		<ul style="list-style-type: none">• Clinical diagnosis of plasma cell myeloma/multiple myeloma AND• Results of bone marrow biopsy unknown or unavailable	<ol style="list-style-type: none">1. Primary site bone marrow (C421)2. Histology plasma cell myeloma/multiple myeloma (9732/3)



Notes / Examples

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

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



Rule	Histology	Other	Code
PH8		Diagnosis is <ul style="list-style-type: none">• Smoldering myeloma• Indolent myeloma• Evolving myeloma• Plasma cell myeloma• Multiple myeloma	1.Primary site bone marrow (C421) 2.Histology plasma cell myeloma/ multiple myeloma (9732/3)

Notes / Examples

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

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

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

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



**Module 3: Lymphoma/leukemia
(Specific neoplasms that can
manifest as either leukemia or
lymphoma)**

PH9-PH12



Module 3 Header cont'd

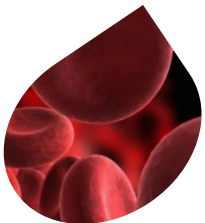
BCCLL/SLL 9823/3

**Blastic plasmacytoid dendritic cell neoplasm,
NOS 9727/3**

Burkitt cell leukemia 9826/3

Burkitt lymphoma, NOS 9687/3

**Precursor B-cell lymphoblastic
leukemia/lymphoma 9836/3**





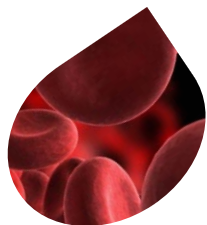
Module 3 Header cont'd

**Precursor B-cell lymphoblastic lymphoma, NOS
9728/3**

**Precursor T-cell lymphoblastic lymphoma, NOS
9729/3**

Small B lymphocytic lymphoma 9670/3

T lymphoblastic leukemia/lymphoma 9670/3



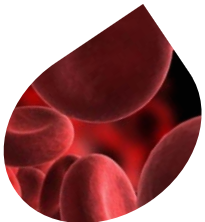


Notes

Note 1: ICD-9-CM and ICD-10 have separate codes for leukemia and lymphoma

Note 2: Commonly lymphoma originates in lymph node region(s), tissue, or organ(s) although it will metastasize to the bone marrow when the disease is stage IV or disseminated

Note 3: Commonly leukemia originates in the bone marrow





Rule	Histology	Other	Code
PH9		<ul style="list-style-type: none">• Diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (BCCLL/SLL) AND• There is peripheral blood involvement (bone marrow may also be involved)	<ol style="list-style-type: none">1. Primary site bone marrow (C421)2. Histology BCCLL/SLL (9823/3)



Notes / Examples

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

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

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



Rule	Histology	Other	Code
PH10	B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma	<ul style="list-style-type: none">• Diagnosis is B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma AND• Peripheral blood and flow cytometry are negative or unknown AND• Cannot verify that disease originated in bone marrow	<ol style="list-style-type: none">1. Primary site to the site of origin (lymph node region(s), tissue, or organ)2. Histology B-cell lymphocytic lymphoma (9670/3)



Notes / Examples

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

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

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

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



Rule	Histology	Other	Code
PH11		Diagnosis is <ul style="list-style-type: none">• Burkitt lymphoma/leukemia OR• Precursor cell lymphoblastic lymphoma/leukemia OR• Precursor B-cell lymphoblastic leukemia/lymphoma OR• Precursor T-cell lymphoblastic leukemia/lymphoma AND• Only involvement is bone marrow	1. Primary site bone marrow (C421) 2. Histology <ul style="list-style-type: none">• Burkitt cell leukemia (9826/3)• Precursor cell lymphoblastic leukemia, NOS (9835/3)• Precursor B-cell lymphoblastic leukemia (9836/3)• Precursor T-cell lymphoblastic leukemia (9837/3)



Notes / Examples

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

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

Rule	Histology	Other	Code
PH12		Diagnosis is <ul style="list-style-type: none"> • Burkitt lymphoma/leukemia OR • Precursor cell lymphoblastic lymphoma/leukemia OR • Precursor B-cell lymphoblastic leukemia/lymphoma OR • Precursor T-cell lymphoblastic leukemia/lymphoma AND • Involvement of lymph node region(s), tissue or organ(s) 	<ol style="list-style-type: none"> 1. Primary site to the site of origin (lymph node region(s), tissue, or organ) 2. Histology <ul style="list-style-type: none"> • Burkitt lymphoma, NOS (9687/3) • Precursor cell lymphoblastic lymphoma, NOS (9727/3) • Precursor B-cell lymphoblastic lymphoma (9728/3) • Precursor T-cell lymphoblastic lymphoma (9729/3)



Notes / Examples

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

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

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

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



Module 4: Preleukemia, Smoldering Leukemia, and Myelodysplastic Syndrome (9989/3) PH13



Rule	Histology	Other	Code
PH13		Diagnosis is <ul style="list-style-type: none">• Preleukemia OR• Smoldering leukemia OR• Myelodysplastic syndrome	<ol style="list-style-type: none">1. Primary site bone marrow (C421)2. Histology myelodysplastic syndrome (9989/3)



Notes / Examples

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



Conclusion

- The new hematopoietic and lymphoid neoplasm rules go into effect for cases diagnosed **January 1, 2010, and after**
- Email address for questions askseerctr@imsweb.com

