Hematopoietic and Lymphoid Neoplasm Project
Primary Site and Histology Rules

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PH Rules

• Rules apply to
  • Problematic sites
  • Problematic 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.
Note 4

Apply the 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 that Module.
Module 5: Myeloid Neoplasms

Histology: 9861/3, 9930/3

Rules: PH14-PH15
Module 5: Rule PH14

Code the primary site bone marrow (C421) and code the histology 9861/3 when the diagnosis is myeloid neoplasm or acute myeloid leukemia, NOS AND the involvement is limited to bone marrow.
Module 5: Rule PH14

**Note:** Do **not** change primary site code because the spleen is involved with infiltrate. The infiltrate refers to deposits of leukemia in the spleen as a result of the spleen filtering the blood.
Module 5: Rule PH15

Code the primary site to the site of origin (lymph node region(s), tissue, or organ) and the code the histology to myeloid sarcoma (9930/3) when the diagnosis is myeloid neoplasm or myeloid sarcoma AND the neoplasm originates in a site other than bone marrow.
Module 5: Rule PH15

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

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

## Table C1: Lymph Node/Lymph Node Chain Reference Table

<table>
<thead>
<tr>
<th>Lymph Node/Lymph Node Chain</th>
<th>ICD-O-3 Code</th>
<th>ICD-O-3 Lymph Node Region(s)</th>
<th>AJCC Lymph Node Region(s)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal</td>
<td>C772</td>
<td>Intra-abdominal</td>
<td>Pelvic, right and left*</td>
</tr>
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<td>C772</td>
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<td>C773</td>
<td>Axilla or arm</td>
<td>Axillary, right and left*</td>
</tr>
<tr>
<td>Anterior cecal</td>
<td>C772</td>
<td>Intra-abdominal</td>
<td>Para-aortic</td>
</tr>
<tr>
<td>Anterior deep cervical</td>
<td>C770</td>
<td>Head, face and neck</td>
<td>Cervical, right and left*</td>
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<td>C772</td>
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<td>C775</td>
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Module 5: *Endnotes*

- For Rules on coding primary site for lymphomas go to Modules 1 and 7.

- When this Module does not apply to the case being abstracted, go to Module 8.
Module 6: Coding Primary Site and Histology for Specified Lymphoma

Histology: 9596/3, 9597/3, 9671/3, 9680/3, 9690/3 9691/3, 9695/3, 9698/3, 9761/3

Rules: PH16-PH24
Module 6: Rule PH16

Code the primary site to the site of origin (lymph node region(s), tissue, or organ) and code the histology diffuse large B-cell lymphoma (DLBCL) (9680/3) when DLBCL (9680/3) and follicular lymphoma (9690/3) are present in the same lymph node(s), tissue, or organ
Module 6: Rule PH16

Note 1: The original pathology may identify only DLBCL although both DLBCL and follicular lymphoma are present. The DLBCL is much more aggressive than the follicular lymphoma and often masks the follicular lymphoma during the initial work-up.
Because it is more aggressive, the DLBCL will respond more rapidly to treatment so the post-treatment biopsies may show a combination of DLBCL and follicular lymphoma or the post-treatment biopsy may be positive for only follicular lymphoma.
Module 6: Rule PH16

The follicular lymphoma was present from the beginning but was hidden. Do **not** change the histology; it should remain 9680/3
Note 2: Do **not** simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Modules 1 and 7 for more information on coding primary site for lymphoma.

Note 3: See Appendix C for help in identifying lymph node names, chains, and codes.
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Module 6: Rule PH16

**Note 4:** Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.
Module 6: Rule PH17

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

- **Example 1:** Diffuse follicular lymphoma, grade 1. Code follicular lymphoma, grade 1 (9695/3)
Module 6: Rule PH17

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

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

• **Example 4:** Follicular lymphoma, diffuse. Code follicular lymphoma, NOS (9690/3).
Module 6: Rule PH18

Code the primary site to skin (C44_) and the histology to follicle cell lymphoma (9597/3) when there is skin infiltration with follicle cell lymphoma or B-cell lymphoma, follicle type and the involvement is limited to skin, or skin and the regional lymph nodes.
Module 6: Rule PH18

**Note:** If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do not code follicle cell lymphoma and do not code skin as the primary site. Dissemination to other sites or distant lymph nodes is uncommon and would occur late in the stage of the disease.
Module 6: Rule PH19

Code the primary site to **skin** (C44_) and the histology to **large B-cell lymphoma** (9680/3) when there is **skin infiltration** with **large B-cell lymphoma** or **B-cell lymphoma, large cell type** and the involvement is limited to **skin**, or **skin** and the **regional lymph nodes**.
Module 6: Rule PH19

*Note:* If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do not code skin as the primary site.
Module 6: Rule PH20

Code the primary site to skin (C44_) and the histology to B-cell lymphoma, NOS (9680/3) when there is skin infiltration with B-cell lymphoma and the involvement is limited to skin, or skin and the regional lymph nodes.
Module 6: Rule PH20

*Note:* If there is involvement of lymph nodes that are not regional for the skin site involved, or involvement of bone marrow or organ(s), do not code skin as the primary site.
Module 6: Rule PH21

Code the primary site to the **site of origin** (lymph node region(s), tissue, or organ) and the histology **composite lymphoma (9596/3)** when **both** non-Hodgkin lymphoma and Hodgkin lymphoma are **present in the same lymph node** region(s), tissue, or organ.
Module 6: Rule PH21

**Note 1:** Use composite lymphoma code when

- Both NHL and HL are present in one lymph node or multiple lymph nodes in one lymph node region.
- Both NHL and HL are present in multiple lymph nodes in one lymph node region or several lymph node regions as defined by ICD-O-3, e.g. NHL and HL present in superior hilum and superior rectal lymph nodes.
- Assume all lymph nodes are involved with both NHL and HL even when only one lymph node is biopsied.
Module 6: Rule PH21

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

**Note 3:** See Appendix C for help in identifying lymph node names, chains, and codes.
Module 6: Rule PH21

**Note 4:** Commonly lymphomas originate in lymph nodes, tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.
Module 6: Rule PH21

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

- NHL is present in one lymph node region and HL is present in another lymph node region, e.g. NHL in cervical lymph node(s) and HL in inguinal lymph node(s)
- NHL in liver and HL in intra-thoracic lymph nodes
Module 6: Rule PH22

Code the primary site to the site of origin (lymph node region(s), tissue, or organ) and the histology to the numerically highest ICD-O-3 code when two or more non-Hodgkin lymphomas are present in the same lymph node(s), tissue, or organ.
Module 6: Rule PH22

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

**Note 2:** See Appendix C for help in identifying lymph node names, chains, and codes.
Module 6: Rule PH22

**Note 3:** Commonly lymphomas originate in lymph node region(s), tissue, or organ(s) although they will metastasize to the bone marrow when the disease is stage IV/disseminated. If nodes, tissue, or organs are involved at the time of diagnosis, code as a lymphoma.
Module 6: Rule PH22

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

- Thymic extranodal marginal-zone B-cell lymphoma is present in the thymus and diffuse large B-cell lymphoma in the hilar lymph nodes.
- B-cell lymphoma is present in the intrathoracic lymph nodes and peripheral T-cell NHL in the liver.
Module 6: Rule PH23

Code the primary site blood (C420) and the histology Waldenstrom macroglobulinemia (9761/3) when there is lymphoplasmacytic lymphoma in the bone marrow and IgM monoclonal gammopathy in the blood.
Module 6: Rule PH24

Code the primary site to the involved bone marrow, lymph nodes, or lymphoid tissue and the histology lymphoplasmacytic lymphoma (9671/3) when the diagnosis is Waldenstrom macroglobulinemia OR lymphoplasmacytic lymphoma and Waldenstrom macroglobulinemia AND the bone marrow, lymph nodes OR lymphoid tissue are involved.
Module 6: *Endnotes*

- For additional Rules on coding primary site for lymphomas go to Modules 1 and 7.

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