Collecting Cancer Data: Hematopoietic Disease

November 4, 2011

NAACCR Cancer Registry & Surveillance Webinar Series

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Questions

- Please use the Q&A panel to submit your questions
- Send questions to "All Panelist"

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Fabulous Prizes!!

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Agenda

- Overview
- Hematopoietic Manual
- Staging
- Treatment

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Overview

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General Terms

- Pluripotential hematopoietic stem cells (PHSC)
  - The precursor cells which give rise to all the blood cell types of both the myeloid and lymphoid lineages.

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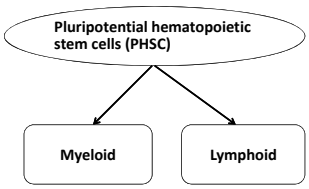
General Terms



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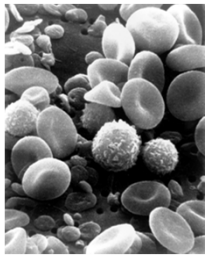
General Terms



- There are three major types of lymphocytes.
  - T-Cells
  - B-Cells
  - Natural Killer (NK) Cells

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T-Cells and B-Cells

- Immature lymphocytes that travel to the thymus differentiate into T-Cells
  - "T" is for thymus
- Immature lymphocytes that travel to the spleen or lymph nodes differentiate into B cells
  - "B" stands for the bursa of Fabricius, which is an organ unique to birds, where B cells mature.

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Diagnostic Confirmation

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

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Diagnostic Process for Leukemia

- Clinical symptoms
  - Weight loss
  - Weakness
  - Bruising
- CBC and/or peripheral blood smear
  - Identifies abnormal white or red blood cells
- Bone marrow biopsy

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Leukemia vs. Lymphoma

- Leukemia most commonly presents in the bone marrow and/or blood
- Lymphoma most commonly manifests in lymph nodes, lymphoid tissue, or lymphoid organs

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Transformation

- Transformation from a chronic to an acute leukemia is a progression of disease
  - Does not compare to the progression of disease in solid tumors
- Histology actually changes when the chronic phase of leukemia progresses or transforms to the acute phase
  - Treatment and survival for chronic and acute leukemia is vastly different

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Hodgkin Lymphoma

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

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Non-Hodgkin Lymphoma

- Non-Hodgkin lymphoma (NHL)
  - Comprises a diverse group of malignant neoplasms which include all lymphomas other than Hodgkin
  - Arises in lymphocytes
  - Commonly develops in lymph nodes but also occurs in extranodal sites

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Lymphatic Organs

- Lymph nodes
- Tonsils
  - Pharyngeal tonsils (adenoids) C11.1
  - Palatine tonsils C09.9
  - Lingual tonsils C02.4
  - Waldeyer ring C14.2
- Spleen
- Thymus
- Peyer's patches

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Lymphoma and Acquired Immunodeficiency Syndrome (AIDS)

- AIDS defining lymphoma
  - Burkitt's lymphoma
  - Immunoblastic lymphoma
  - Primary central nervous system (CNS) lymphoma
- AIDS related lymphoma
  - Hodgkin lymphoma
  - Non-Hodgkin lymphoma

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2010 Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual

Effective with Cases Diagnosed 1/1/2010 and after

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What's New in Hematopoietic & Lymphoid Neoplasm Data Collection

- *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues, 4th Edition*
  - Reference for rules and information in database
- Changes in reportability
  - New ICD-O histology terms and codes
  - Existing codes change from borderline (/1) to malignant (/3) behavior
  - Transformations collected as new primaries

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What's New in Hematopoietic & Lymphoid Neoplasm Data Collection

- Use the *Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual* and the Hematopoietic Database for cases diagnosed 2010 and later
  - Manual and Database replace:
    - *Single Versus Subsequent Primaries of Lymphatic and Hematopoietic Diseases* table (February 2001)
    - *Abstracting and Coding Guide for the Hematopoietic Diseases (2002)*
    - Previous casefinding and Reportable Neoplasm lists for hematopoietic neoplasms (ICD-9-CM and ICD-10)

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What's New in Hematopoietic & Lymphoid Neoplasm Data Collection

- Use the *Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual* and the Hematopoietic Database for cases diagnosed 2010 and later
  - Manual and Database provide:
    - Case reportability instructions
    - Multiple primary rules
    - Primary site and histology coding rules
    - Grade coding rules

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Case Reportability Instructions

1. Report case when only information is that cancer-directed treatment* for a reportable hematopoietic or lymphoid neoplasm has started
2. Report the case when the hematopoietic or lymphoid neoplasm diagnosis is preceded by an ambiguous term

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*http://www.nci.nih.gov/cancertopics/pdq/cancerdatabase

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Ambiguous Terms that Constitute a Diagnosis of Hematopoietic or Lymphoid Neoplasm

- Apparent(ly)
- 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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Case Reportability Instructions

3. Report case when there is a clinical diagnosis of reportable hematopoietic or lymphoid neoplasm
4. Report case when multiple myeloma, evolving myeloma, early multiple myeloma, indolent multiple myeloma or smoldering multiple myeloma is diagnosed
5. Report case when preleukemia or smoldering leukemia is diagnosed

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Case Reportability Instructions

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

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Case Reportability Instructions

7. Report case when a reportable diagnosis appears in any text or report described as definitive diagnostic method in Hematopoietic Database
8. Report hematopoietic and lymphoid neoplasms with ICD-O-3 morphology codes 9590-9992 that are listed as /1 and described as malignant by a physician

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Case Reportability Instructions

9. Report all ICD-O-3 morphology codes 9590-9992 with a /3 behavior plus the new histology terms and codes published by *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues, 4th Edition* (See Appendix D)
10. Query the Hematopoietic Database to determine case reportability for cases that do not meet criteria listed in previous instructions (1-9)

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Multiple Primary Rules

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Notes

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

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Multiple Primaries

- Rule M1
  - Abstract as a single primary when minimal information is available (such as a death certificate only (DCO) case or a pathology-report-only case).
- Rule M2
  - Abstract as a single primary when there is a single histology.

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Multiple Primaries

- Rule M3
  - Abstract as a single primary when two or more types of non-Hodgkin lymphoma are present in the same anatomic location(s), such as one lymph node, one organ, or one tissue.
- Rule M4
  - Abstract as a single primary when both Hodgkin and non-Hodgkin lymphoma are present in the same anatomic location(s). Hodgkin and non-Hodgkin may be present in one lymph node, one organ, or one tissue

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Multiple Primaries

- Rule M5
  - Abstract as multiple primaries when any of the following situations are met
    - Hodgkin lymphoma in one node and non-Hodgkin lymphoma in a different node
    - Hodgkin lymphoma in one organ and non-Hodgkin lymphoma in a different organ
    - Hodgkin lymphoma in tissue and non-Hodgkin lymphoma in different tissue

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Multiple Primaries

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

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Multiple Primaries

- Rule M7
  - Abstract as a single primary when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days AND there is documentation of one positive bone marrow biopsy
- Rule M8
  - Abstract as multiple primaries when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days AND there is documentation of two bone marrow examinations, one confirming the chronic neoplasm and another confirming the acute neoplasm

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Multiple Primaries

- Rule M9
  - Abstract as a single primary when both the chronic and the acute phase of the neoplasm are diagnosed within 21 days AND there is no available documentation on bone marrow biopsy
- Rule M10
  - Abstract as multiple primaries when a neoplasm is originally diagnosed in a chronic (less aggressive) phase AND second diagnosis of a blast or acute phase more than 21 days after the chronic diagnosis

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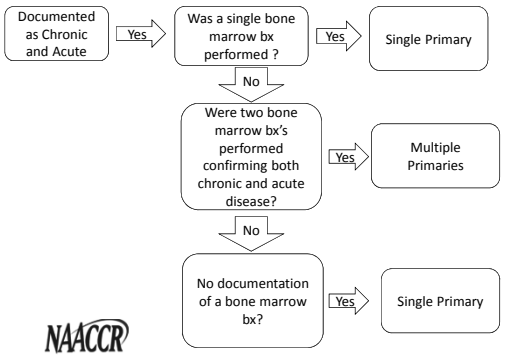
Chronic/Acute Diagnosed within 21 days



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Multiple Primaries

- Rule M11
  - Abstract the acute phase as a single primary when a neoplasm is originally diagnosed in the blast or acute phase and reverts to a less aggressive/chronic phase and there is no confirmation available that the patient has been treated.
- Rule M12
  - Abstract as multiple primaries when a neoplasm is originally diagnosed in the blast or acute phase and reverts to a less aggressive/chronic phase after treatment.

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Multiple Primaries

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

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Pop Quiz

- Patient had two cervical lymph nodes excised.
  - One contained Nodular sclerosis classical Hodgkin lymphoma.
  - The other contained Burkitt lymphoma.
- How many primaries?

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Pop Quiz

- Patient presents with a history of follicular lymphoma, nos. Now has a lymph node biopsy that shows malignant large cleaved follicular lymphoma.
- How many primaries?

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Primary Site and Histology Coding Rules

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Modules 1-9

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

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Module 1: General Instructions

- Rule PH1
  - Code the primary site using information from scans, documentation in the medical record, the pathology report, and from the Hematopoietic DB.
- Rule PH2
  - Code the histology diagnosed by the definitive diagnostic method(s) (see Hematopoietic DB).
- Rule PH3
  - Code the primary site and histology using the medical practitioner's statement on the medical record or death certificate when none of the tests or reports defined as a definitive diagnostic method is available.

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Module 2: Plasma Cell Neoplasms

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

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Module 2: Plasma Cell Neoplasms

- Rule PH5
  - Code the primary site to the specific bone (C400-C419) where the plasmacytoma originated and code the histology solitary plasmacytoma of bone (9731/3) when the diagnosis is
    - Plasma cell neoplasm
    - Solitary plasmacytoma
    - Solitary plasmacytoma of bone
    - Solitary medullary plasmacytoma
    - Multiple plasmacytomas
    - Multiple plasmacytomas of bone
    - Multiple medullary plasmacytomas

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Module 2: Plasma Cell Neoplasms

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

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Module 2: Plasma Cell Neoplasms

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

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Module 2: Plasma Cell Neoplasms

- Rule PH8
  - Code the primary site bone marrow (C421) and the histology plasma cell myeloma/multiple myeloma (9732/3) when the diagnosis is smoldering myeloma, indolent myeloma, evolving myeloma, plasma cell myeloma, or multiple myeloma.
- When this module does not apply to the case being abstracted, go to Module 8.

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Module 3: Lymphoma/Leukemia

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

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Module 3: Lymphoma/Leukemia

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

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Module 3: Lymphoma/Leukemia

- Rule PH11
  - Code the primary site bone marrow (C421) and the respective histology from the list below when the diagnosis is Burkitt lymphoma/leukemia, precursor cell lymphoblastic lymphoma/leukemia, precursor B-cell lymphoblastic leukemia/lymphoma, or precursor T-cell lymphoblastic leukemia/lymphoma AND the only involvement is bone marrow.
    - Burkitt cell leukemia (9826/3)
    - Precursor cell lymphoblastic leukemia, NOS (9835/3)
    - Precursor B-cell lymphoblastic leukemia/lymphoma (9836/3)
    - T lymphoblastic leukemia/lymphoma (9837/3)

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Module 3: Lymphoma/Leukemia

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

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Module 3: Lymphoma/Leukemia

- PH12 (cont)
  - Burkitt lymphoma, NOS (9687/3)
  - Blastic plasmacytoid dendritic cell neoplasm (previously called precursor cell lymphoblastic lymphoma, NOS) (9727/3)
  - Precursor B-cell lymphoblastic lymphoma, NOS (9728/3)
  - Precursor T-cell lymphoblastic lymphoma, NOS (9729/3)
- **When this module does not apply to the case being abstracted, go to Module 8.**

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Module 4: Preleukemia, Smoldering leukemia and Myelodysplastic syndrome

- Rule PH13
  - Code the primary site bone marrow (C421) and the histology myelodysplastic syndrome (9989/3) when the diagnosis is preleukemia, smoldering leukemia, or myelodysplastic syndrome.
- **When this module does not apply to the case being abstracted, go to Module 8.**

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Module 5: Myeloid Neoplasms

- 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
- Rule PH15
  - Code the primary site to the site of origin (lymph node region(s), tissue, or organ and the histology to myeloid sarcoma (9930/3) when the diagnosis is myeloid neoplasm or myeloid sarcoma AND the neoplasm originates in a site other than bone marrow.
- **When this module does not apply to the case being abstracted, go to Module 8.**

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

- 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

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

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

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

- Rule PH18
  - Code the primary site to skin (C44_) and the histology to follicle cell lymphoma (9597/3) when there is skin infiltration with follicle cell lymphoma or B-cell lymphoma, follicle type and the involvement is limited to skin, or limited to skin and the regional lymph nodes.
- Rule PH19
  - Code the primary site to skin (C44_) and the histology to large B-cell lymphoma (9680/3) when there is skin infiltration with large B-cell lymphoma or B-cell lymphoma, large cell type and the involvement is limited to skin, or limited to skin and the regional lymph nodes.

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

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

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

- 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.

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

- 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.

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Module 6: Coding Primary Site and Histology for Specified Lymphoma

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

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Module 7: Primary Site Rules for Lymphomas Only

- Rule PH25
  - Code the primary site to the specific lymph node region when only one lymph node or one lymph node region is involved.
- Rule PH26
  - Code the primary site mediastinal lymph nodes (C771) when the site of lymphoma is described only as a mediastinal mass.
- Rule PH27
  - Code the primary site intra-abdominal lymph nodes (C772) when the site of lymphoma is described only as a retroperitoneal mass or as a mesenteric mass.

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Module 7: Primary Site Rules for Lymphomas Only

- Rule PH28
  - Code the primary site inguinal lymph nodes (C774) when the site of lymphoma is described only as an inguinal mass.
- Rule PH29
  - Code the primary site to the specific lymph node region when multiple lymph node chains within the same region (as defined by ICD-O-3) are involved.

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Module 7: Primary Site Rules for Lymphomas Only

- Rule PH30
  - Code the primary site as multiple lymph node regions, NOS when multiple lymph node regions (C778) as defined by ICD-O-3 are involved and it is not possible to identify the lymph node region where the lymphoma originated.
- Rule PH31
  - Code the primary site to lymph nodes, NOS (C779) when lymph node(s) are involved but no primary site/particular lymph node region is identified.

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Module 7: Primary Site Rules for Lymphomas Only

- Rule PH32
  - Code the primary site to bone marrow (C421) when lymphoma is present only in the bone marrow.
- Rule PH33
  - Code the primary site to the specific organ when lymphoma is present only in an organ.

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Module 7: Primary Site Rules for Lymphomas Only

- Rule PH34
  - Code the primary site to the lymph node region as defined by ICD-O-3 when there is proof of extension from the regional lymph nodes into the organ.
- Rule PH35
  - Code the primary site to the organ when lymphoma is present in an organ and that organ's regional lymph nodes.

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Module 7: Primary Site Rules for Lymphomas Only

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

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Module 7: Primary Site Rules for Lymphomas Only

- Rule PH37
  - Code primary site to unknown primary site (C809) only when there is no evidence of lymphoma in lymph nodes AND the physician documents in the medical record that he/she suspects that the lymphoma originates in an organ(s). See ICD-O-3 Rule D.
- **When this module does not apply to the case being abstracted, go to Module 8.**

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Module 8: Histology Rules Only
All hematopoietic and lymphoid neoplasms

- Rule PH38
  - Code the non-specific (NOS) histology when the diagnosis is:
    - One non-specific histology AND
    - Two or more specific histologies AND
    - The hematopoietic DB multiple primaries calculator documents the specific histologies and NOS are the same primary AND
    - No further information is available

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Module 8: Histology Rules Only
All hematopoietic and lymphoid neoplasms

- Rule PH39
  - Code the specific histology when the diagnosis is:
    - One non-specific (NOS) histology AND
    - One specific histology AND
    - The Hematopoietic DB multiple primaries calculator documents the specific histology and NOS are the same primary
- **When this module does not apply to the case being abstracted, go to Module 9.**

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Module 9: Default Rules
All hematopoietic and lymphoid neoplasms

- Rule PH40
  - Use the Hematopoietic DB to determine the primary site and histology when rules PH1-PH39 do not apply.
- Rule PH41
  - When the histology code cannot be determined using the Hematopoietic DB, code the histology with the numerically higher ICD-O-3 code.

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Pop Quiz

- A lymph node biopsy shows B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (BCCLL/SLL). A peripheral blood smear shows the same.
  - What is the primary site?
  - What is the histology?

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Grade

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Grade

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

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Grade-code 9

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

These neoplasms do not have a specific codable phenotype

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Grade

- Rule G2
  - Use statements from any part of the medical record including, but not limited to
    - Pathology report OR
    - History and physical OR
    - Consultation OR
    - Final diagnosis OR
    - Face sheet

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Grade

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

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Grade

- Rule G4
  - Code T-cell (code 5) for the neoplasms listed to the right.
- Rule G5
  - Code T-cell (code 5) when the neoplasm is identified
    - T-cell
    - T-cell phenotype
    - T-precursor
    - Pre-T
    - gamma-delta-T
    - null-cell T-cell

- 9701/3
- 9702/3
- 9705/3
- 9708/3
- 9709/3
- 9714/3*
- 9716/3
- 9717/3
- 9718/3
- 9724/3
- 9725/3
- 9726/3
- 9827/3
- 9831/3
- 9834/3
- 9837/3

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* Unless the pathologist specifically designates as a B-cell

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Grade

- Rule G60
  - Code B-cell (code 6) for the B-cell precursor lymphoid neoplasms and the mature B-cell neoplasms listed on the right.

- 9591/3
- 9596/3
- 9597/3
- 9670/3
- 9671/3
- 9673/3
- 9678/3
- 9679/3
- 9680/3
- 9684/3
- 9687/3
- 9688/3
- 9689/3
- 9690/3
- 9691/3
- 9695/3
- 9698/3
- 9699/3
- 9712/3
- 9728/3
- 9731/3
- 9732/3
- 9734/3
- 9737/3
- 9738/3
- 9762/3
- 9811/3
- 9812/3
- 9813/3
- 9814/3
- 9815/3
- 9816/3
- 9817/3
- 9818/3
- 9823/3
- 9833/3
- 9836/3
- 9940/3

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Grade

- Rule G7
  - Code B-cell (code 6) when the disease is identified as B-cell, B-cell phenotype, B-precursor, pre-B, or null-cell and B-cell.
- Rule G8
  - Code Null cell, non-T non-B (code 7) when the disease is described as null cell, non-T non-B, or common cell.

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Grade

- Rule G9 Code NK-cell (natural killer cell) (code 8) for
  - Extranodal NK/T cell lymphoma, nasal type 9719/3
  - Aggressive NK-cell leukemia 9948/3
    - Aggressive NK-cell leukemia/lymphoma
    - Large granular cell lymphocyte leukemia
    - NK-cell type REAL
- Rule G10
  - Code Natural Killer (NK) cell (code 8) when the disease is described as NK cell, natural killer cell, nasal NK/T-cell lymphoma, or null-cell and NK cell.

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Grade

- Rule G11
  - Code cell type not determined, not stated, not applicable (code 9) when
  - There is no statement describing the cell type OR
  - The cell type is described as combined T AND B cell OR
  - The cell type is described as combined B AND NK cell

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Pop Quiz

- A patient is diagnosed and treated at your facility for Refractory anemia 9980/3.
  - What grade would be assigned to this histology?
  - What rule would be used to assign this code?

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Appendices

- Appendix A
  - History of Hematopoietic and Lymphoid Neoplasm Coding
- Appendix B
  - WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues Histology Lineage
- Appendix C
  - Lymph Node/Lymph Node Chain Reference Table

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Appendices

- Appendix D
  - New Histology Terms and Codes Hematopoietic and Lymphoid Neoplasms
- Appendix E
  - Histology "NOS" Tables
- Appendix F
  - Master Code Lists

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Questions?

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CSv2
Hematopoietic Disease
HemeRetic Schema

Slides have been adapted from the CSv2 Training Materials

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CS Extension

Code	Description
100	Localized disease: single, solitary, unifocal, isolated, mono-ostotic Plasmacytomas (9731/3) Extramedullary plasmacytoma (9734/3) Mast cell sarcoma (9740/3) Malignant histiocytosis (9750) Histiocytic sarcoma (9755) Langerhans cell sarcoma (9756/3) Dendritic cell sarcoma (9757/3, 9758/3) Myeloid sarcoma (9930/3) Langerhans cell histiocytosis (9751/3)
800	Systemic disease (poly-ostotic): All histologies including those in 100
999	Unknown

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Remaining CS Data Items

- All these fields are "not applicable"
  - CS Tumor Size = 988
  - CS Eval fields (TS/Ext, Reg Nodes, Mets) = 9
  - CS Lymph Nodes = 988
  - Reg LN Pos and Reg LN Exam = 99
  - CS Mets at DX = 98

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Site-specific Factor 1: JAK-2

Code	Description
000	JAK-2 test result stated as negative
010	JAK2 test performed, positive for mutation V617F in exon 14
020	JAK2 test performed, positive for mutation of exon 12
800	JAK2 test performed, positive for other specified mutation
810	JAK2 test performed, positive for more than one mutation
850	JAK2 test performed, positive NOS; specific mutation(s) not stated
888	Obsolete
988	Not applicable
997	Test ordered, results not in chart
998	Test not done
999	Unknown

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New Data Items

Data Item	Code	Explanation
Grade Path Value	Blank	Not applicable
Grade Path System	Blank	Not applicable
Lymph Vascular Invasion	8	Not applicable
Mets at DX – Bone	8	Not applicable
Mets at DX – Lung	8	Not applicable
Mets at DX – Brain	8	Not applicable
Mets at DX – Liver	8	Not applicable

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CSv2 Lymphoma

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Lymphatic Sites

- Lymph nodes (C77._)
- Thymus (C37.9)
- Spleen (C42.2)
- Peyer's patches (C17.2)
- Nodules in the Appendix (C18.1)
- Waldeyer's Ring (C14.2)
  - Pharyngeal tonsil
  - Tubal tonsils
  - Palatine tonsils
  - Lingual tonsils

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Extralymphatic Sites

- Stomach
- Small intestine
- Gastrointestinal tract
- Brain
- Lung
- Any organ can be involved by lymphoma

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The Diaphragm

- The diaphragm's role in staging:
  - Muscle that separates the chest (thoracic) cavity from the abdomen
  - Dividing point for staging in lymphomas (below and above, on both sides)
- The diaphragm is used in several ways:
  - Same side of the diaphragm
  - Opposite side of the diaphragm
  - Both sides of the diaphragm

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CS Extension - Lymphatic

- 100 (Stage I)
  - Primary site: Lymphatic
  - Involvement of single lymph node region
- 200 (Stage II)
  - Primary site: Lymphatic
  - Two or more lymph node regions involved on same side of diaphragm
- 300 (Stage III)
  - Primary site: Lymphatic
  - Lymph node regions involvement on both sides of the diaphragm

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CS Extension-Extralymphatic

- 110 (Stage IE)
  - Primary site: Extralymphatic organ
  - Involvement of extralymphatic organ only
- 210 (Stage IIE)
  - Primary site: Extralymphatic organ
  - Lymph node involvement on same side of diaphragm
- 310 (Stage IIIE)
  - Primary site: Extralymphatic organ
  - Lymph node involvement on both sides of diaphragm

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CS Extension - Spleen

- 120 (Stage IS)
  - Primary site: Spleen ONLY
- 220 (Stage IIS)
  - Primary site: Spleen or lymphatic structure
  - Spleen involved
  - Lymph nodes below the diaphragm involved
- 230 (Stage IIES)
  - Primary site: Extralymphatic organ
  - Spleen involved
  - Extralymphatic organ below the diaphragm involved
  - Lymph nodes below the diaphragm involved/not involved

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CS Extension - Spleen

- 320 (Stage IIIS)
  - Primary site: Spleen or lymphatic structure
  - Spleen involved
  - Lymph nodes above diaphragm or on both sides of diaphragm involved
- 330 (Stage IIIES)
  - Primary site: Extralymphatic organ
  - Spleen involved
  - Lymph nodes involved on opposite or both sides of the diaphragm

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CS Extension

- 800 (Stage IV)
  - Lymphatic sites (including Spleen)
    - Metastatic sites (Bone marrow, Liver, Lung)
  - Extralymphatic sites
    - Diffuse or disseminated (multifocal) involvement of one or more extralymphatic sites with or without lymph node involvement
    - Involvement of isolated extralymphatic organ in absence of involvement of adjacent lymph nodes, but in conjunction with disease in distant sites
    - Metastatic sites (Bone marrow, Liver, Lung)
- 999 (Unknown)

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CS Extension Summary Table

Nodal/ Lymphatic	Extranodal/ Extralymphatic	Spleen (involvement)
100	110	120*
200	210	220, 230
300	310	320, 330
800	800	800
999	999	999

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CS Tumor Size (TS)/Ext Eval

- Clinical Staging (TS/Ext Eval = 0 or 9)
  - Use code 0 when biopsies, CT's, MRI's or other imaging source determines extension
  - Use 0 when the primary site is surgically removed for an extranodal site
    - Example: Tonsillectomy for tonsillar lymphoma
  - Use code 9 (unknown) when there is no workup or you don't know how staging information was obtained
    - (i.e. Physician states lymphoma is Stage I, no other information available)

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CS TS/Ext Eval

- Pathological staging (TS/Ext eval = 3)
  - Staging laparotomy REQUIRED, which includes
    - Laparotomy/laparoscopy
    - Splenectomy
    - Liver, lymph node & bone marrow biopsies
- Autopsy staging (TS/Ext eval = 2 or 8)
  - Code 2 added to match all other sites
    - No surgical resection done, but evidence derived from autopsy (tumor was suspected or diagnosed prior to autopsy)
  - Code 8
    - Evidence from autopsy only (tumor was unsuspected or undiagnosed prior to autopsy)

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Remaining CS Data Items

- All these fields are "not applicable"
  - CS Tumor Size = 988
  - CS Eval fields (Reg Nodes, Mets) = 9
  - CS Lymph Nodes = 988
  - Reg LN Pos and Reg LN Exam = 99
  - CS Mets at DX = 98

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Site-Specific Factor 1
Associated with HIV/AIDS

Code	Description
001	Yes/Present
002	No/Not present
999	Unknown

•If there is no mention of AIDS in the chart and no evidence of HIV testing, code 999 for unknown

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Site-Specific Factor 2
Systemic Symptoms at Diagnosis

Code	Description
000	No B symptoms
010	Any B symptoms: Night sweats Unexplained fever (above 38 degrees C) Unexplained weight loss (generally greater than 10% of body weight in the six months before admission) B symptoms, NOS
020	Pruritis (if recurrent and unexplained)
030	(010) + (020)
999	Unknown

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Site-specific Factors 3-5
Coding Guidelines

- If index or score is named (IPI, FLIPI, IPS) and point value is given, code score in the appropriate site-specific factor and code other two SSFs as 999
- If index or score is not named and a point value of 5 or less is documented, use code 999 in all three SSFs
- If score is 6 or 7, assume that it is IPI
  - Code score in SSF5
  - Code SSFs 3 and 4 as 999

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Site-specific Factors 3-5

Coding Guidelines

- If risk is stated as "low," "intermediate," or "high" but index or score is not named, use code 999 in all three SSFs
- If score is named and both point value and risk category are documented, code point value
- Code only the statement/score/index documented by the clinician
- Do not calculate score or risk category based on medical record information

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Site-Specific Factor 3

International Prognostic Index (IPI)

Code	Description
000	0 points
001	1 point
002	2 points
003	3 points
004	4 points
005	5 points
990	Stated as low risk (0-1 points)
991	Stated as low intermediate risk (2 points)
992	Stated as intermediate risk (3 points)
993	Stated as high risk (4-5 points)
999	Unknown

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Site-Specific Factor 4

Follicular Lymphoma Prognostic Index (FLIPI)

Code	Description
000	0 points
001	1 point
002	2 points
003	3 points
004	4 points
005	5 points
888	Obsolete
988	Not applicable
990	Stated as low risk (0-1 points)
991	Stated as intermediate risk (2 points)
992	Stated as high risk (3-5 points)
999	Unknown

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Site-Specific Factor 5
International Prognostic Score (IPS)

Code	Description
000	0 points
001	1 point
002	2 points
003	3 points
004	4 points
005	5 points
006	6 points
007	7 points
888	Obsolete
988	Not applicable
999	Unknown

112

New Data Items

Data Item	Code	Explanation
Grade Path Value	Blank	Not applicable
Grade Path System	Blank	Not applicable
Lymph Vascular Invasion	8	Not applicable
Mets at DX – Bone	8	Not applicable
Mets at DX – Lung	8	Not applicable
Mets at DX – Brain	8	Not applicable
Mets at DX – Liver	8	Not applicable

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Treatment

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Treatment

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

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Treatment

- Induction therapy
  - Initial treatment of a patient with a blood cancer with chemotherapy (or radiation therapy)
  - Aim is to kill a maximum number of blood cancer cells to induce a remission (absence of signs or effects of the disease)

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Treatment

- Consolidation therapy
  - Treatment of acute leukemia with drug treatment given to patients in remission after induction therapy.
  - Aim is to kill as many of the remaining cancer cells as possible

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Surgery

- Excisional biopsy of a lymph node
  - If the intent of the procedure was diagnostic, assign code 02 under Diagnostic Staging Procedure
  - If the intent was treatment, use code 25 for Surgical Procedure of Primary Site

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Question

- If a patient receives a stand alone procedure to assess lymph node involvement of a tumor, no other tissue/cells are removed do we code assessment of the lymph node always to scope of regional lymph node surgery?

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Answer

- For solid tumors, code the lymph node aspiration, biopsy, or resection in the Scope of Regional Lymph Node field.
- If a lymph node is biopsied or removed to diagnose or stage lymphoma, and that node is NOT the only node involved with lymphoma, use code 02 in Surgical Diagnostic and Staging Procedure field . If that node resected is the only node diagnosed with lymphoma, use the Surgical Procedure of the Primary Site code for the lymphoma primary - 25.

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4/29/2010

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Radiation Therapy
Radiation Treatment Volume

- 01-60: Specific site
  - 22: Mantle
  - 23: Lower extended field
    - Middle (para-aortic) field
    - Pelvic field
  - 39: Inverted Y
    - Middle and pelvic fields
  - 60: Lymph node region, NOS

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Systemic Therapy

- Primary treatment for most hematopoietic and lymphoid neoplasms
  - Varies widely based on diagnosis

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Systemic Therapies

- Rituximab
- CHOP
- ABVD
- Anthracycline

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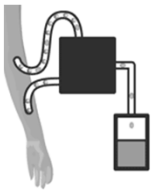
Hematologic Transplant

- Bone marrow transplant
  - Code 11: autologous
  - Code 12: allogenic
- Peripheral blood stem cell transplant
  - Code 20



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http://www.lymphoma-net.org/transplantation.cfm

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Thank YOU!!!

- Next Month's webinar
  - Collecting Cancer Data: Liver and Biliary Tract
    - 12/2/2010

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