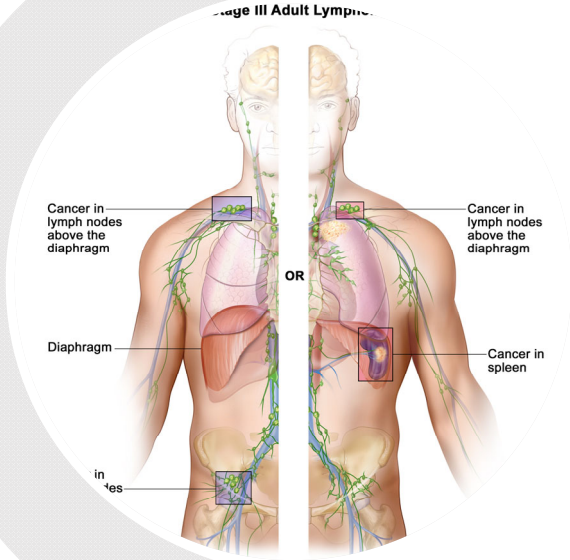


LYMPHOMA
2021

NAACCR 2020-2021 WEBINAR SERIES

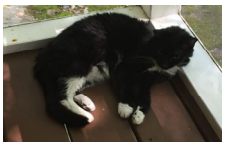
Q&A

- Please submit all questions concerning the webinar content through the Q&A panel.
- If you have participants watching this webinar at your site, please collect their names and emails.
- We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.

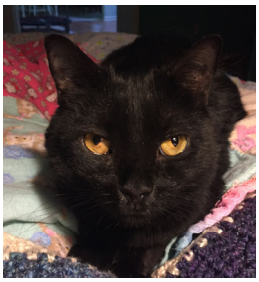
FABULOUS PRIZES

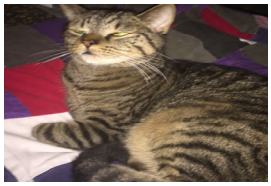
3

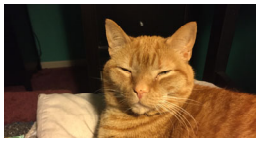
GUEST PRESENTER



- Jennifer Ruhl, et al.
  - Chair SSDI WG
  - Public Health Analyst NIH/NCI SEER

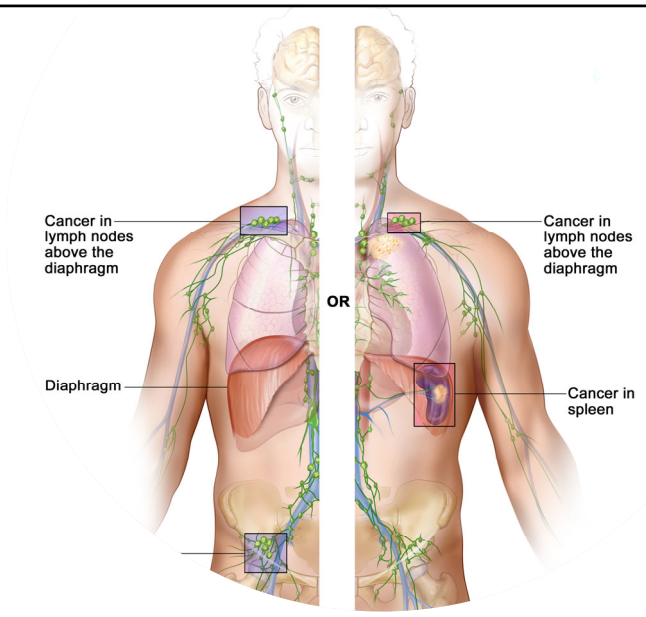






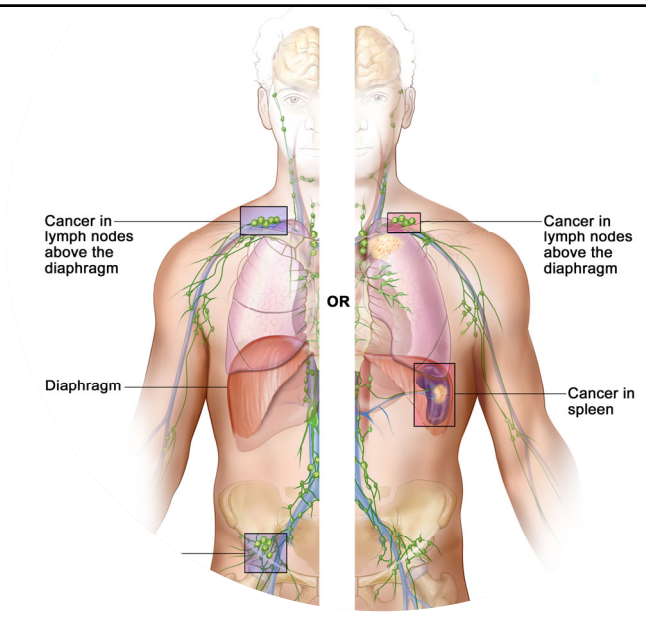
AGENDA

- Anatomy/Staging
- Hematopoietic Database/Manual
  - New histologies (Effective for 1/1/2021+)
  - Changes in reportable for existing histologies (Effective for 1/1/2021+)
  - CLL/SLL
  - Multiple primary rules
  - Primary Site and Histology (PH) Rules
  - Other Data Items and Hematopoietic Neoplasms (supplemental)
- Case Scenarios



STAGING

- Overview
- AJCC
- Summary Stage
- EOD



LYMPHOMA (EXCLUDING CLL/SLL)

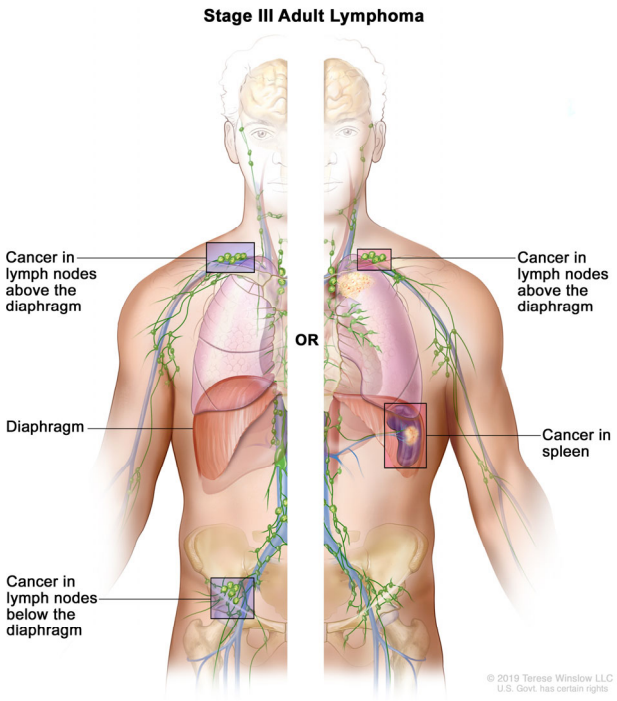
- AJCC Chapters 79 Hodgkin and Non-Hodgkin and 80 Pediatric Hodgkin and Non-Hodgkin
- Summary Stage 2018 Chapter: Lymphoma
- SSDI's
  - Schema Discriminator for 9591/3
  - B symptoms
  - HIV status
  - NCCN International Prognostic Index (IPI)

LYMPHOMA-CLL/SLL

- AJCC Chapters 79 and 80
- Histology 9823
- Lugano Stage used for AJCC Stage
- Summary Stage 2018 Chapter: Lymphoma
- SSDI's (in addition to SSDIs in Lymphoma): Used to determine RAI stage
  - Adenopathy
  - Anemia
  - Lymphocytosis
  - Organomegaly
  - Thrombocytopenia

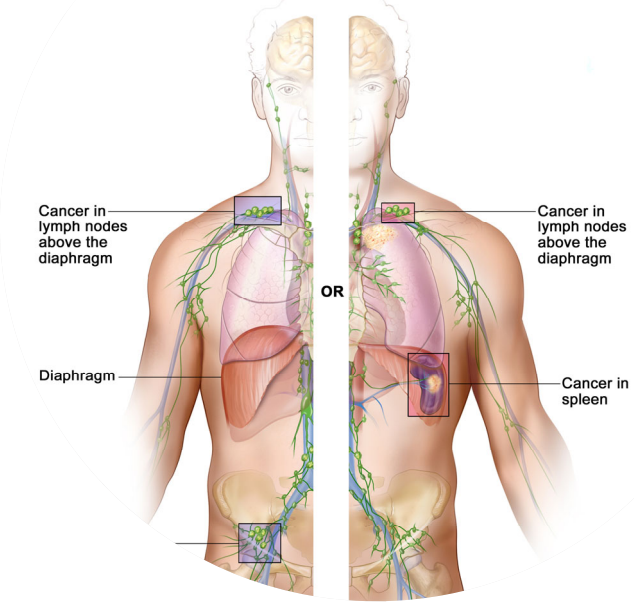
AJCC STAGE CHAPTER REVIEW

Chapter 79 Hodgkin and Non-Hodgkin Lymphomas



LUGANO CLASSIFICATION FOR HODGKIN AND NON-HODGKIN LYMPHOMA

- This is the stage that is recorded in the AJCC Stage Group data item.
  - Used for all lymphomas eligible for staging in chapter 79 (including CLL/SLL).



10

EXTRANODAL DISEASE (E)

- Waldeyer's ring, thymus, spleen are considered nodal.
  - Do not use E suffix
- The (E) suffix is used for lymphomas that arise in extranodal sites or when lymphoma arising from a node extends into an extranodal site.
  - Liver is an exception. Any liver involvement is Stage 4.
  - The (E) suffix may only be used with Stage 1 or Stage 2 disease. It is no longer valid with Stage 3 disease.

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BULKY DISEASE

- Indicates a clinically enlarged mass
  - Hodgkin Lymphoma (HL)
    - If mediastinal, "Bulky" is defined as greater than 1/3 the size of the cavity.
    - If not mediastinal, "Bulky" is defined as greater than 10cm
  - Non-Hodgkin Lymphoma (NHL)
    - Definition varies based on histology.
  - Look for physician statement of "Bulky"
  - Stage 2 Bulky is a new stage category for 8th edition

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A/B CLASSIFICATION

- Relevant for Hodgkin Lymphoma
- No longer included as part of stage group
- Collected as an SSDI

13

CLL/SLL

- Lugano Classification for Hodgkin and Non-Hodgkin lymphoma
  - This is the stage information collected in the *AJCC TNM Clin Stage Group* data item.
  - Involvement of peripheral blood alone is **NOT** stage 4
    - This is relevant for all lymphomas, but peripheral blood involvement only is mostly seen with CLL/SLL
- Modified Rai staging system and Binet staging system
  - **Not** collected in the *AJCC TNM Clin Stage Group* data item.
  - Components of the Rai and Binet system collected in SSDI's.

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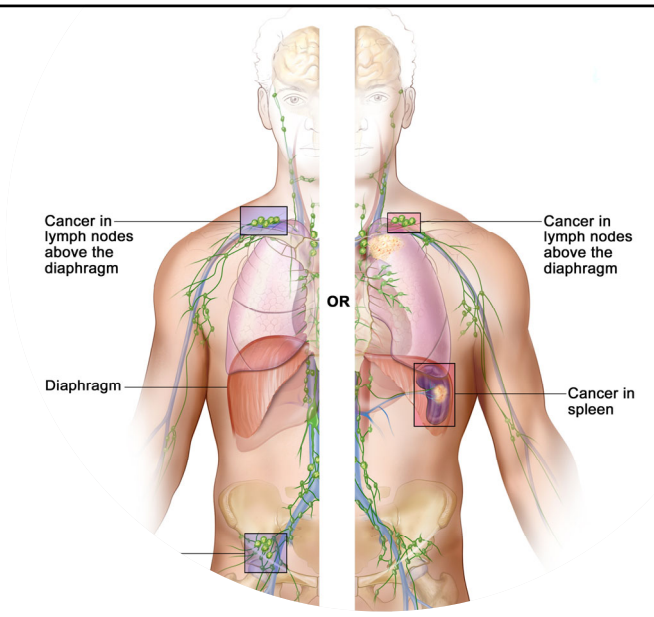
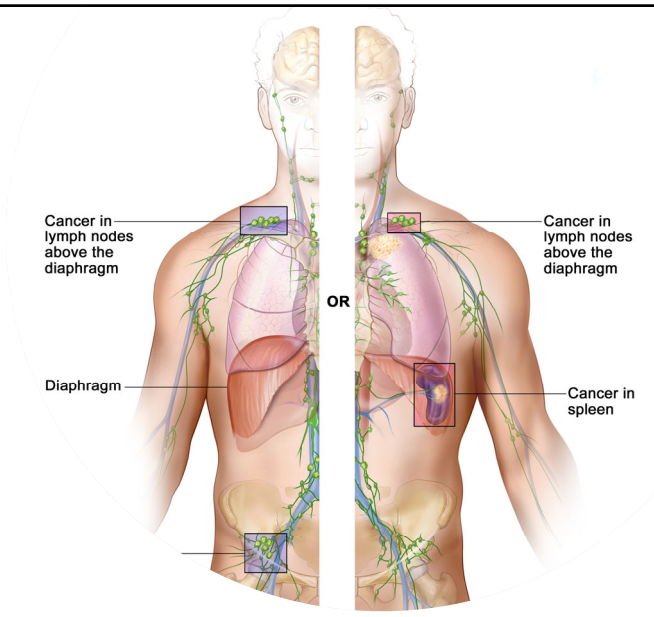
LUGANO

- Is there a single lymph node or single lymph node chain involved?
- Is there a single extralymphatic site involved (not bulky disease)?



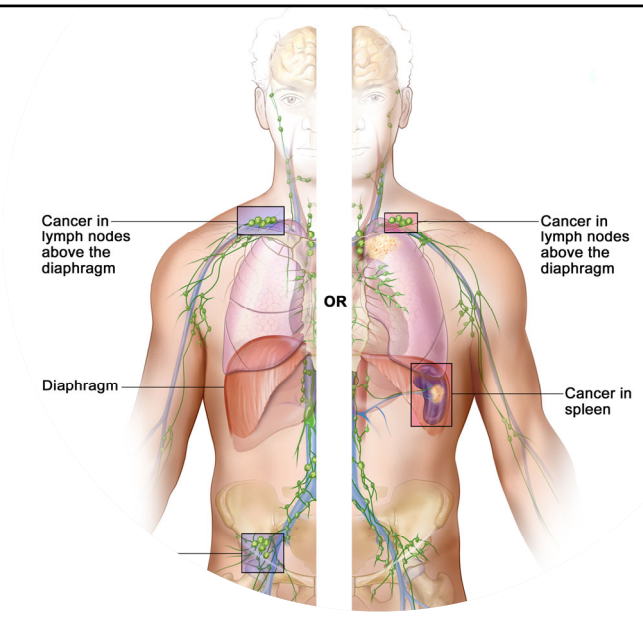
LUGANO

- Are there multiple lymph node chains involved on the same side of the diaphragm?
- Is there a single extra-lymphatic site and regional nodes?
- Is there direct extension from a lymph node into an extranodal site?
- Does the patient have “Bulky” disease



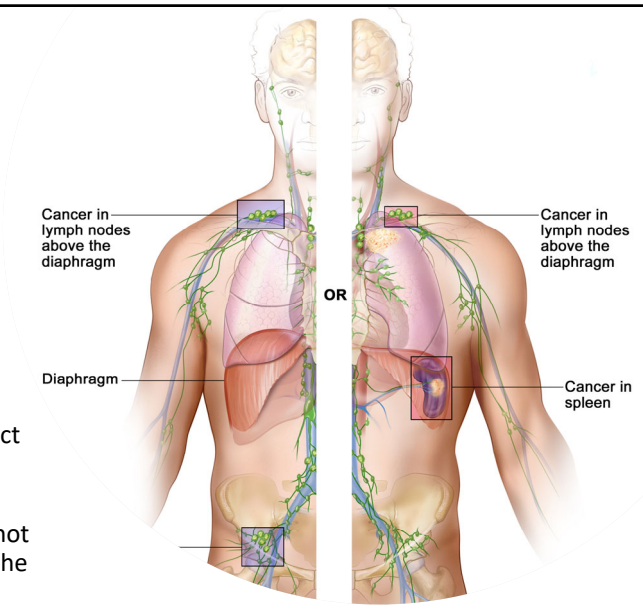
LUGANO

- Are lymph node regions above and below the diaphragm involved?
- Are lymph nodes above the diaphragm and the spleen involved?



LUGANO

- Is there involvement of cerebral spinal fluid (CNF)?
- Is there involvement of bone marrow?
- Is there liver involvement?
- Are there multiple lung lesions?
- Is there diffuse involvement of an extralymphatic organ(s)?
- Is there extralymphatic involvement (excluding direct extension) with multiple lymph node region involvement?
- Is there extra lymphatic involvement and direct or not direct) and lymph nodes involved on both sides of the diaphragm?



SUMMARY STAGE 2018

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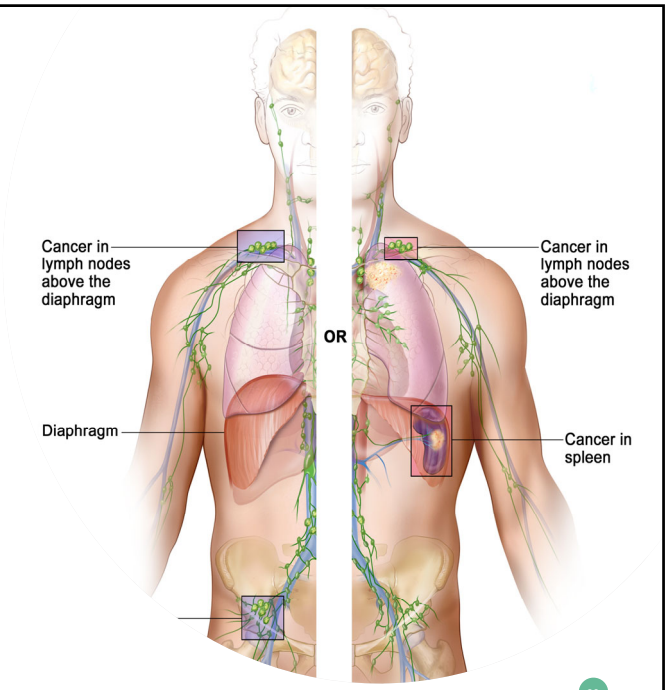
LYMPHOMA

- Code 1: Localized: single nodal chain involved or single organ involved
- Code 2: Previously code 5 (SS2000)
  - Bulky disease present
  - Involvement of regional nodes on same side diaphragm



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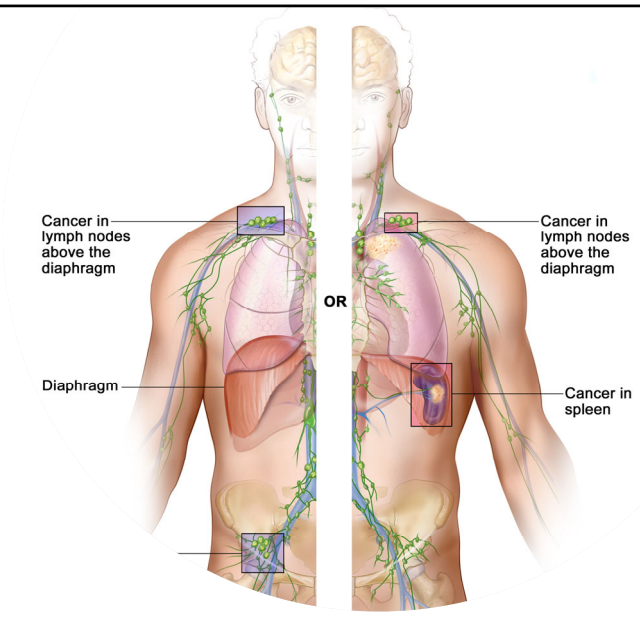
LYMPHOMA

- Code 7:
  - Involvement of regional LNs on both sides of diaphragm
  - Two or more extralymphatic sites involved
  - Diffuse or disseminated involvement of extralymphatic sites
  - Distant metastasis: bone marrow, CSF, CNS, Lung



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EOD

Primary Tumor

Regional Nodes

Metastasis

22

LYMPHOMAS

- EOD Primary Tumor
  - Code 100: Nodal lymphoma confined to one LN chain
  - Code 200: Extranodal lymphoma confined to site
  - Code 300: Nodal lymphoma with two or more LN chains involved on SAME side of diaphragm
  - Code 400: Extranodal lymphoma with involvement of regional lymph nodes

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LYMPHOMAS

- EOD Primary Tumor
  - Code 500: Codes 300 or 400 WITH bulky disease
  - Code 600: Involvement of lymph nodes on both sides of the diaphragm
  - Code 700: Diffuse or disseminated (multifocal) involvement of ONE OR MORE extralymphatic organ(s)/site(s)
    - WITH or WITHOUT lymph node involvement

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LYMPHOMAS

- EOD Primary Tumor
  - Code 750: Peripheral Blood Involvement only (mostly seen with CLL/SLL)
  - Code 800: Metastatic involvement
    - Bone marrow
    - Cerebrospinal fluid (CSF)
    - CNS (if not CNS primary)
    - Lung (if not Lung primary)

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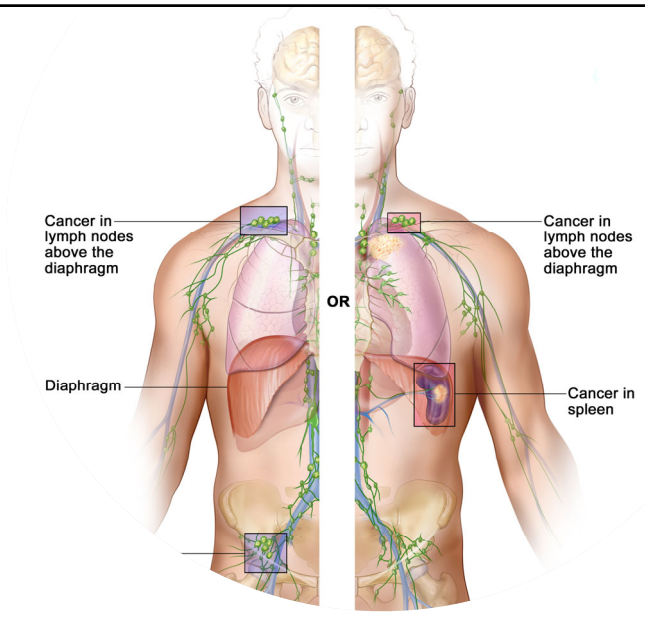
QUESTIONS?



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Hematopoietic and Lymphoid Neoplasms

Jennifer Ruhl, MSHCA, RHIT, CCS, CTR
NCI, Public Health Analyst

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12/16/2020

New Histologies

Effective for 1/1/2021+

New Histologies with a /1 (Effective 1/1/2021+)

- **9680/1: EBV-positive mucocutaneous ulcer**
  - EBVMCU; EBV+ mucocutaneous ulcer
  - May *transform* to DLBCL (9680/3). The DLBCL would be the first reportable primary
- **9738/1: HHV8-positive germinotopic lymphoproliferative disorder**
  - GLPD; Usually occurs in HIV-negative individuals; Involves the lymph nodes
  - Reportable form HHV8-positive diffuse large B-cell lymphoma (9738/3)
- **9823/1: Monoclonal B-cell lymphocytosis, CLL-type**
  - MBL, atypical CLL-type; MBL, chronic lymphocytic leukemia (CLL)-type; MBL, CLL-type; MLUS; Monoclonal B lymphocytosis of uncertain significance
  - May *transform* to CLL/SLL (9823/3). The CLL/SLL would be the first reportable primary

In Situ Lymphomas (Effective 1/1/2021+)

- Per Hematopoietic Manual, Case Reportability Instructions #3: *Do **NOT** report in situ (/2) lymphomas*
- **9673/1: In situ mantle cell neoplasia**
  - In situ mantle cell lymphoma, ISMCN, Mantle cell lymphoma-like B cells of uncertain/undetermined significance
- **9695/1: In situ follicular neoplasia**
  - Follicular lymphoma in situ, In situ follicular lymphoma, Intrafollicular neoplasia, ISFN

New Histology (Effective 1/1/2021+)

- **9715/3: Anaplastic large cell lymphoma, ALK-negative**
  - ALK-ALCL
  - Breast implant-associated ALCL
    - Primary site C50_ (C509 if quadrant can't be determined)
  - Seroma-associated ALCL
- Term currently reportable and coded as 9702/3
  - For cases with diagnoses less than 1/1/2021 with ALK-, code to 9702/3
- *Note: Edits will not allow this histology to be used prior to 1/1/2021*

New Histology (Effective 1/1/2021+)

- **9749/3: Erdheim-Chester disease (ECD)**
  - Clonal systemic proliferation of histiocytes
  - Commonly having a foamy (xanthomatous) component, and contain Touton giant cells (type of non-Langerhans cell histiocytosis)
  - This neoplasm is not reportable prior to 2021
- *Note: Edits will not allow this histology to be used prior to 1/1/2021*

New Histology (Effective 1/1/2021+)

- **9766/3: Lymphomatoid granulomatosis, grade 3**
  - Term currently reportable and coded as 9680/3
  - For cases with diagnoses less than 1/1/2021 with Lymphomatoid granulomatosis, grade 3, code to 9680/3

Note: Edits will not allow this histology to be used prior to 1/1/2021

- Note: the following are coded as 9766/1
  - Lymphoid granulomatosis, NOS (no grade specified)
  - Lymphoid granulomatosis grade 1 or 2

New Histology (Effective 1/1/2021+)

- **9819/3: B-lymphoblastic leukemia/lymphoma, BCR-ABL1-like**
  - Lacks BCR-ABL1 translocation, shows pattern of gene expression very similar to that seen in ALL with BCR-ABL1
- Term currently reportable and coded as 9811/3
  - For cases with diagnoses less than 1/1/2021 with this diagnosis, code 9811/3
- *Note: Edits will not allow this histology to be used prior to 1/1/2021*

New AML Histologies (Effective 1/1/2021+)

- **9877/3: Acute myeloid leukemia with mutated *NPM1***
- **9878/3: Acute myeloid leukemia with biallelic mutation of CEBPA**
- **9879/3: Acute myeloid leukemia with mutated *RUNX1***
- **9912/3: Acute myeloid leukemia with *BCR-ABL1***

- Terms currently reportable and coded as 9861/3
  - For cases with diagnoses less than 1/1/2021 with this diagnosis, code 9861/3

- *Note: Edits will not allow these histologies to be used prior to 1/1/2021*

New Histologies (Effective 1/1/2021+)

- **9968/3: Myeloid/lymphoid neoplasms with *PCM2-JAK2***
  - Term currently reportable and coded as 9975/3
  - For cases with diagnoses less than 1/1/2021 with this diagnosis, code 9975/3

- **9993/3: Myelodysplastic syndrome with ring sideroblasts and multilineage dysplas**ia
  - Term currently reportable and coded as 9985/3
  - For cases with diagnoses less than 1/1/2021 with this diagnosis, code 9985

- *Note: Edits will not allow these histologies to be used prior to 1/1/2021*

Changes in Existing Histologies

Effective for 1/1/2021

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Alternate names now a /1 (Effective 1/1/2021+)

- **9702/1: Indolent T-cell lymphoproliferative disorder of the gastrointestinal tract**
  - Previously an alternate name for 9702/3 (2001-2020 in Heme DB)
  - Most common in small intestine and colon
  - Lymphoid cells infiltrate lamina propria but usually do not show invasion of the epithelium
  - May transform to a higher-grade T-cell lymphoma, which would be a reportable primary
- **9709/1: Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder**
  - Previously an alternate name for 9709/3 (2001-2020 in Heme DB)
  - Primary cutaneous CD4+ small/medium T-cell lymphoma [OBS]
  - Solitary/multiple plaques or nodules, no evidence of patches

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Changes in behavior (/3 to /1) (Effective 1/1/2021+)

- **9725/3: Hydroa vacciniforme-like lymphoma (very rare)**
  - 9725/1: Hydroa vacciniforme like lymphoproliferative disorder
  - No longer reportable as of 1/1/2021
- **9971/3: Post –transplant lymphoproliferative disorder (PTLD)**
  - 9971/1: Post-transplant lymphoproliferative disorder (PTLD)
  - No longer reportable as of 1/1/2021
  - *Note*: If PTLD is diagnosed with a lymphoma, still report the lymphoma (Rule M14)

Obsolete Histologies and their new code (Effective 1/1/2021+)

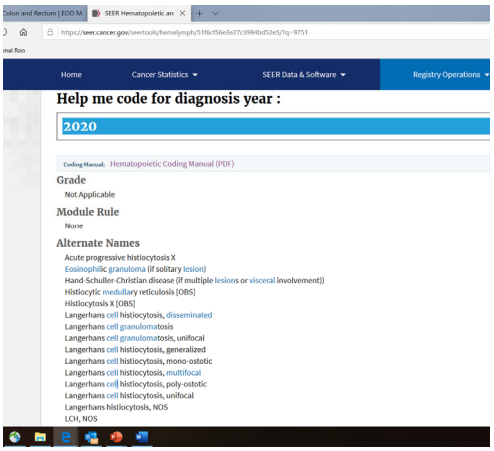
- **9826/3: Burkitt Leukemia**
  - Now coded as 9867/3: Burkitt lymphoma
  - 9826/3 can no longer be used with diagnosis dates 1/1/2021+ (edits enforced)
- **9991/3: Refractory neutropenia**
  - Now 9980/3: Myelodysplastic syndrome with single lineage dysplasia
  - 9991/3 can no longer be used with diagnosis dates 1/1/2021+ (edits enforced)
- **9992/3: Refractory thrombocytopenia**
  - Now 9980/3: Myelodysplastic syndrome with single lineage dysplasia
  - 9992/3 can no longer be used with diagnosis dates 1/1/2021+ (edits enforced)

Langerhans Cell Histiocytosis (9751/3) (Effective 1/1/2021+)

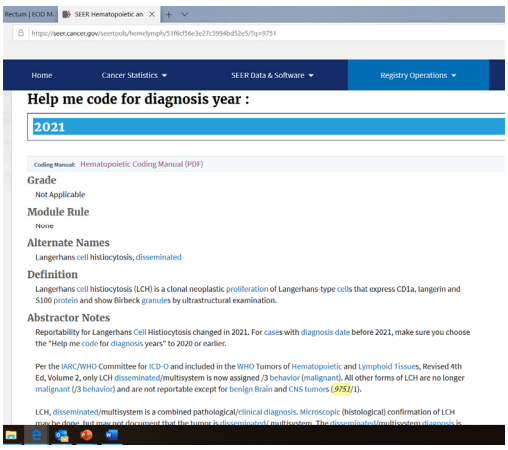
- For 2021+, only Langerhans cell histiocytosis, **disseminated** is a /3
  - All other terminology, including Langerhans cell histiocytosis, NOS is now a /1
- Defining the "disseminated" diagnosis:
  - Is a combined pathological/clinical diagnosis
  - The "disseminated" diagnosis will not be found on the pathology report
  - The disseminated/multisystem diagnosis is based on clinical evaluation by the managing/treating physician and is based on multiple areas of involvement
    - Note: Not all suspected sites of involvement need to be biopsied or surgically resected

Langerhans Cell Histiocytosis (9751/3) and the Hematopoietic Database

Heme Database for 2010-2020 diagnoses



Heme Database for 2021 diagnoses

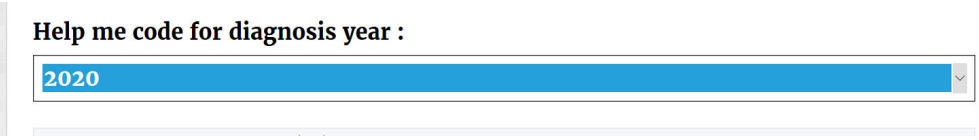


Langerhans Cell Histiocytosis (9751/3) (Effective 1/1/2021+)

- New Abstractor Note
  - Per the IARC/WHO Committee for ICD-O and included in the WHO Tumors of Hematopoietic and Lymphoid Tissues, Revised 4th Ed, Volume 2, only LCH disseminated/multisystem is now assigned /3 behavior (malignant). All other forms of LCH are no longer malignant (/3 behavior) and are not reportable except for benign Brain and CNS tumors (*9751*/1)
- Note: There is now a 9751/1 as well, which includes this note
  - 9751/1 can be used for benign brain tumors in the CNS/Brain/Intracranial Gland

Choosing the Right Year

- Make sure that you have the right year
- Look for “Help me code for diagnosis year”
- Make sure that if you are working on a 2020 or earlier case, that the year chosen is 2020
- If you have a 2021 case, make sure the diagnosis year 2021 is chosen
- This is extremely importantly for
  - New histologies, obsolete histologies, changes in behavior



CLL/SLL-The problem child of Hematopoietic

Diagnosing CLL/SLL (and other lymphomas/leukemias)

- CLL/SLL is frequently diagnosed via **peripheral blood smear**
- Per Diagnostic Confirmation, Code 1, #3
  - Peripheral blood smear
    - Can be used as a histological diagnosis for any of the hematopoietic histologies (9590/3-9992/3)
- Per recent clarification:
  - Peripheral blood smear followed by flow cytometry (most commonly done with CLL/SLL) is Diagnostic confirmation code #3 (See Heme Manual, Code 3: 1c)
    - If unable to find documentation that a peripheral blood smear was done first, assume that it was and code 3
    - The flow cytometry is what is confirming the diagnosis

CLL/SLL (9823/3)

- Per AJCC 8th edition, now **staged** as Lymphoma
  - Does **not** mean that primary sites are C77_ (Lymph nodes)
- Assigning primary site
  - If blood/peripheral blood or bone marrow is involved, primary site is C421 (does not matter what else is involved)
    - See Rule PH5
  - If there is **no or unknown** blood/peripheral blood or marrow involved, assign primary site based on involvement of lymph nodes and/or organs
    - See Rule PH6
    - Could still be a stage IV based on other involvement

CLL/SLL (9823/3)

- **Do not default to primary site C421 (bone marrow) when diagnosis is CLL/SLL**
- Per the Hematopoietic Manual
  - *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*
- This note is telling you that CLL/SLL usually indicates extensive disease
- It is **not** telling you that the bone marrow and/or peripheral blood is always involved, and that primary site is automatically C421

CLL/SLL (9823/3)

- CLL/SLL: Lugano Staging (effective AJCC 8th edition)
  - Lugano is primarily for SLL (usually no bone marrow involvement)
  - T, N, and M are 88
  - Documented as Stages I, IE, 2, 2E, 2 bulky, 3, 4
  - Based on involvement of organ(s), lymph node(s), bone marrow, etc.
  - Very similar to Ann Arbor Stage, which was collected for several years (at least AJCC 6th and 7th editions)
  - If stage is unknown, code 99

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CLL/SLL (9823/3)

- CLL/SLL: RAI Stage, primarily for CLL (bone marrow involvement)
  - This is Stage 0, 1, 2, 3, 4
  - Based on the information collected in the following SSDIs: Adenopathy, Anemia, Lymphocytosis, Organomegaly, Thrombocytopenia
  - No Stage Group for RAI is being assigned/derived at this time
    - Do not record this information in the TNM fields
  - Fill out the SSDIs for the individual elements for the RAI stage
    - SSDI work group working on Derived RAI Stage for implementation in 2021, will be applied to cases diagnosed 2018 and forward
    - When data item implemented, will **not** require additional work for registrars, who will continue to fill out the 5 SSDIs

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Peripheral Blood Involvement

- For Lymphomas: If there is peripheral blood involvement **ONLY**
  - Common for CLL/SLL
  - Primary site C421
  - Per AJCC, this would be **UNSTAGED**
    - AJCC Stage Group 99
    - Clarification received from AJCC recently
    - CAnswer Forum post: http://cancerbulletin.facs.org/forums/forum/ajcc-tnm-staging-8th-edition/hematologic-malignancies-chapters-78-83/hodgkin-and-non-hodgkin-lymphomas-chapter-79/85708-cll-primary-site-c42-1-ajcc-group-stage/page2

Peripheral Blood Involvement: EOD and Summary Stage

- **If there is peripheral blood involvement ONLY**
  - EOD Primary Tumor: Code 750
    - Peripheral Blood Involvement only
      - Can be used for cases diagnosed 2018+ forward, no review of previous cases abstracted are required
  - Summary Stage code 7
    - The recent clarification from AJCC does not change this
- **This only affects Lymphomas (mostly CLL/SLL)**
  - **Reminder: Leukemias are not staged**

Other Lymphomas or Leukemias/Lymphomas (excluding CLL/SLL)

- If peripheral blood/bone marrow is involved, do **NOT** automatically assign primary site C421
  - That rule is for CLL/SLL only
- Review record to see if there is other involvement, such as lymph nodes or organs
  - Peripheral blood/bone marrow involvement would be recorded in stage only when there is lymph node or organ involvement
- For other lymphomas (See Modules 6 & 7) or leukemias/lymphomas (See Module 4), assign primary site C421 when the only involvement is bone marrow and/or peripheral blood

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Multiple Primary (M) Rules

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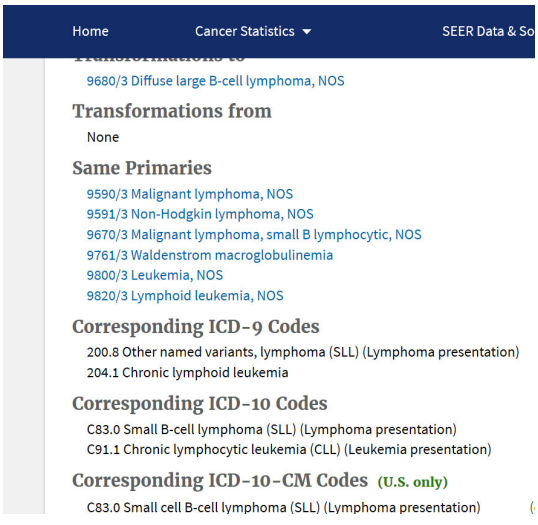
Rule M2: Same histology always same primary

- Reminder: There is no time limit on this
  - This applies to recurrence in any part of the body, or multiple areas of the body
- Same histology is equal to “same primary”
- Same primaries can be found in the Heme DB for each histology

Example of Same Primaries (9823/3: CLL/SLL)



- Snapshot from the Hematopoietic Database
- The same primaries are going to be different depending on the histology
- If you use the Multiple Primaries Calculator with 9823/3 with one of the histologies listed, the MPC will return “same primary”
- *Example*: Originally diagnosed with CLL/SLL. Several years later, bone marrow biopsy done, which came back as lymphoid leukemia, NOS
  - Same primary

M4: Abstract single primary when two or more types of NHL are simultaneously present in the same anatomic location(s)

- Applies to any of the NHL lymphomas
  - Non-Hodgkin Lymphomas are defined as not being Hodgkin Lymphomas
  - Hodgkin Lymphomas are: 9650-9653, 9655, 9659, 9663
- "Simultaneously present in the same anatomic location" means
  - Biopsy of lymph node, organ or bone marrow shows a diagnosis of two lymphomas
- Most common occurrence (per questions in Ask SEER Registrar):
  - DLBCL (9680/3) and the Follicular Lymphomas (9690/3, 9691/3, 9695/3, and 9698/3)

M4: Abstract single primary when two or more types of NHL are simultaneously present in the same anatomic location(s)

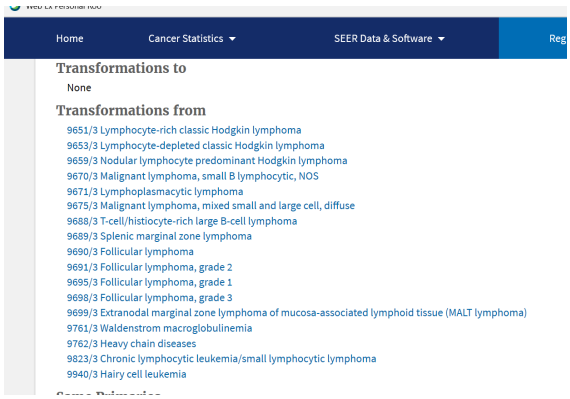
- To assign histology, referred to PH11 and PH15 to assign the histology
  - PH11 states that if DLBCL is one of the histologies, code histology to 9680/3
    - This is because the DLBCL is the more aggressive lymphoma and the patient will be treated for the DLBCL
  - PH15 states to go with the higher histology
- Note: For the DLBCL and Follicular Lymphoma
  - This is also a chronic neoplasm (Follicular lymphoma) and an acute neoplasm (DLBCL)
  - Chronic/Acute rules could also be applied, which would give the same answer

M8-M13 Chronic/Acute Rules

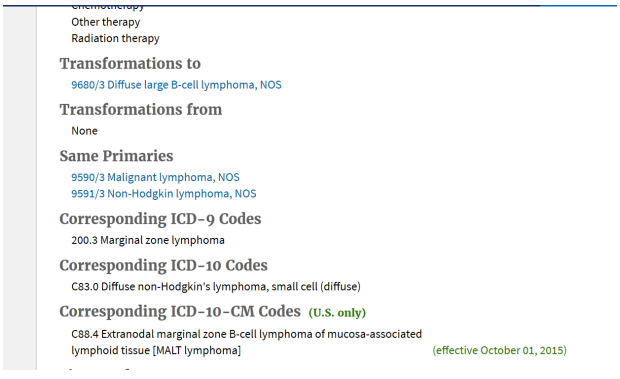
- How do you determine if you have a chronic and an acute neoplasm
- There are two fields in the Hematopoietic Database related to Transformations
  - This is the only place where you will get this information
  - It is not available in the Hematopoietic manual
- Transformations to:
  - This will show the histologies that a neoplasm will “transform to”
  - This means that this is a chronic neoplasm
- Transformations from:
  - This will show the histologies that “transformed from”
  - This means that this is an acute neoplasm

M8-M13 Chronic/Acute Rules

- **9680/3: Example of an Acute Neoplasm**



9699/3: Example of a Chronic Neoplasm



Chronic/Acute Rules: 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)

- This is a common occurrence for the chronic/acute rules
- For this situation, only one biopsy is done which shows both the chronic and the acute neoplasm present in that biopsy
- This is common when a chronic neoplasm is in the process of transforming
- The treatment will be targeted to the acute neoplasm

Chronic/Acute Rules: 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 * Prepare one abstract. Use the primary site and histology coding rules to assign the appropriate primary site and histology codes

- This should be a rare occurrence for the chronic/acute rules
- In this situation, there is no documentation on how or when the two histologies were diagnosed
- Code the later diagnosis
- If you are unable to determine when they were diagnosed, default to the acute

Chronic/Acute Rules: 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
- 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

- This is the most common occurrence for the chronic/acute rules
- Patient diagnosed first with the chronic neoplasm, and then later diagnosed with the acute neoplasm
  - The acute neoplasm could be diagnosed within months or years
- Two abstracts would be completed: one for the chronic, one for the acute

Chronic/Acute Rules: 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 biopsies:** bone marrow , lymph node, or tissue: one confirming the **chronic** neoplasm and another confirming the **acute** neoplasm

- This is a common occurrence for the chronic/acute rules
- Patient diagnosed with biopsy (example: lymph node biopsy) with a chronic neoplasm and than during the same clinical work up, diagnosed with an acute neoplasm on another type of biopsy (example: bone marrow biopsy)
  - The major criteria for this rule is the two separate biopsies

Chronic/Acute Rules: M12 & M13

- These rules cover when an acute neoplasm is diagnosed first, then the chronic is diagnosed later (after the initial workup and treatment plan)
- These are rare occurrences

- Per Rule M12, if there is no treatment for the acute neoplasm, or it is unknown if there is treatment, it is a single primary, the acute neoplasm
- Per Rule 13: If there is documented treatment for the acute neoplasm, then the chronic neoplasm would be abstracted as a second primary

Rule M15: Using the Multiple Primaries Calculator

- **ONLY use the MPC when the rules instruct you to**
  - Misuse of the MPC may give you the wrong number of primaries
  - Based on questions received in Ask a SEER CTR, registrars are using the MPC before going through the rules

Primary Site and Histology (PH) Rules

Primary Site and Histology Coding Rules: PH1

- This rule is primarily for PTLD, which is now a /1
- This rule is still applicable for cases diagnosed 2010-2020
  - Only applicable when PTLD is the only diagnosis for 2010-2020
- As a reminder, a diagnosis of PTLD with an associated lymphoma, or plasmacytoma, or some other reportable neoplasm, is always coded to the lymphoma, plasmacytoma, or other
  - This has not changed

Primary Site and Histology Coding Rules: PH18

- Note indicating that other types of "mass" are not included in this rule, only those that are listed
  - *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*
- Rule only applies to the following
  - Mediastinal Mass (C771)
  - Retroperitoneal or mesenteric mass (C772)
  - Inguinal mass (C774)
  - Pelvic mass (C775)

Module 7 (PH18-PH27)

- Very important note included at the beginning of this module
  - Secondary involvement of distant lymph nodes (for an extranodal lymphoma), bone marrow, liver, multifocal lung, spleen or CNS are included in the stage fields only. This secondary involvement excludes rare primary lymphoid neoplasms of spleen, 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
- Bottom line: Not all sites of involvement are used for assigning primary site. Some sites may be metastatic

Primary Site and Histology Coding Rules: PH21

- Multiple lymph node REGIONS involved and not possible to determine where lymphoma started
- Terms indicating lymph node involvement for nodal lymphomas
  - Enlarged, fixed, lymphadenopathy, matted, palpable, shotty
- Review all available imaging documentation to look for areas of involvement

Primary Site and Histology Coding Rules: PH22

- Organ involvement
- Lymph nodes NOT regional (distant) for that organ involved
- Multiple organs and lymph nodes involved and there is no other information on where lymphoma originated

- Remember: Lymph nodes, C779, take priority when a primary site cannot be determined and there is involvement of lymph nodes and organ(s)

Primary Site and Histology Coding Rules: PH28

- This rule applies when ONLY the bone marrow is involved
  - If any other organ, tissue or lymph nodes are involved, then bone marrow involvement is secondary/distant involvement and bone marrow is NOT the primary site
    - *Exception* for CLL/SLL (9823/3)-See Rules PH3 & PH4
  - When bone marrow biopsy is diagnostic of lymphoma, before assigning primary site of bone marrow
    - Check ALL imaging to see if there is evidence of lymph node involvement

Other Data Items and Hematopoietic Neoplasms

Tumor Size: Clinical, Pathological, Summary

- Code 999 (not applicable) for the following
  - Any case coded to primary site C420, C421, C423, C424, C770-C779, or C809
  - 00830 HemeRetic (excluding Spleen, C422)
  - 00458 Kaposi Sarcoma
  - 00790: Lymphoma (excluding CLL/SLL)
  - 00795: Lymphoma-CLL/SLL
  - 00671 & 00672: Melanoma Iris, Melanoma Choroid and Ciliary Body
  - 00821: Plasma Cell Myeloma
  - 00822: Plasma Cell Disorders
  - 00830: HemeRetic

Mets at Dx Fields (Bone, Brain, Liver, Lung, Distant Lymph Nodes, Other)

- Major change for 2021 for Hematopoietic Neoplasms
  - Some of the histologies included in the HemeRetic schema are eligible for the Mets at Diagnosis fields
    - Dendritic neoplasms (9756/3-9759/3)
    - Erdheim-Chester Disease (9749/3-new histology for 2021)
    - Langerhans cell histiocytosis, disseminated (9751/3)
    - Mast cell sarcoma (9740/3)
    - Myeloid Sarcoma (9930/3)

Mets at Dx Fields (Bone, Brain, Liver, Lung, Distant Lymph Nodes, Other)

- Historically these neoplasms have never been coded as having metastatic disease; however, we would like to start collecting this information if available
- For those collecting EOD
  - EOD Regional Nodes and EOD Mets are coded to not applicable; however, you still need to code the Mets at Dx fields
  - If EOD Primary Tumor is coded to 100 (Localized), then there are no Mets
    - Code all Mets at Dx field as 0
  - If EOD Primary Tumor is coded to 700 (Systemic disease), then there are Mets

Mets at Dx Fields

- If primary site is Bone Marrow (C421), do not record this in Mets at Dx Other. This is the primary site, not a metastatic site
- For other lymphomas where primary site is NOT bone marrow (C421) and bone marrow is involved
  - Code Mets at Dx Other-1 (includes bone marrow)
  - Do not code bone marrow involvement in Mets at Dx-Bone

Surgery of Primary

- If only one lymph node is involved (no additional disease is identified), code Surgery of Primary site 25. Use Lymph Node surgery codes.
  - 25 Local tumor excision, NOS
- If more than one lymph node is involved, assign an excisional biopsy of a lymph node as a Diagnostic Staging Procedure code 02
  - 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. If there is only a single lymph node involved with lymphoma, use the data item *Surgical Procedure of Primary Site* [1290] to code these procedures
    - STORE Manual Surgical Diagnostic and Staging Procedure bullet 4

Surgery of Primary Site/Surgical Margins/Reason for No Surgery-Updated Instructions

- SEER Manual, Surgery of Primary Site, Note #10
- Code 98 for the following primary sites unless the case is death certificate only
  - Any case coded to C420, C421, C423, C424, C760-C768, or C809
- SEER Manual, Surgery of Primary Site, Note #11
  - When Surgery of Primary Site is coded 98
    - Code Surgical Margins of the Primary Site to 9
    - Code Reason for No Surgery of Primary Site to 1
- Same instructions have been incorporated into the STORE Manual

Surgery of Primary Site/Surgical Margins/Reason for No Surgery-Updated Instructions

- Once again, instructions for Hematopoietic neoplasms have been updated
- Surgery of primary site can now be coded for several HemeRetic neoplasms
  - Dendritic neoplasms (9756/3-9759/3)
  - Erdheim-Chester Disease (9749/3-new histology for 2021)
  - Langerhans cell histiocytosis, disseminated (9751/3)
  - Mast cell sarcoma (9740/3)
  - Myeloid Sarcoma (9930/3)
  - Plasmacytomas (9731/3, 9734/3)

Scope of Regional Lymph Node Surgery-Updated Instructions

- Assign code 9 for
  - Any case coded to primary site: C420, C421, C423, C424, C589, C700-C709, C710-C729, C751-C753, C761-C768, C770-C779, or C809
  - 00790: Lymphoma (excluding CLL/SLL)
  - 00795: Lymphoma-CLL/SLL
  - 00822: Plasma Cell Disorder

Scope of Regional Lymph Node Surgery-Updated Instructions

- Scope of Regional Lymph Node Surgery can now be coded for:
  - Dendritic neoplasms (9756/3-9759/3)
  - Erdheim-Chester Disease (9749/3-new histology for 2021)
  - Langerhans cell histiocytosis, disseminated (9751/3)
  - Mast cell sarcoma (9740/3)
  - Myeloid Sarcoma (9930/3)
  - Plasmacytomas (9731/3, 9734/3)
- For those collecting EOD
  - EOD Lymph nodes is still coded to 888

Regional Nodes Examined/Positive

- Code 99 for the following cases:
  - Any case coded to primary site C420, C421, C423, C424, C589, C700-C709, C710-C729, C751-C753, C761-C768, C770-C779, or C809
  - 00790: Lymphoma (excluding CLL/SLL)
  - 00795: Lymphoma-CLL/SLL
- As noted in Scope of Regional Lymph Node Surgery, there are now some HemeRetic histologies that can have regional nodes examined/positive coded

Submit a Question to a SEER Registrar

Questions submitted through this form will be sent to the appropriate SEER personnel. The question and answer may be added to the SEER Inquiry System for others to reference.

Choose a subject

Reporting Guidelines

- ○ Hematopoietic Rules (database and manual)
- ○ Multiple Primary & Histology Rules (for cases diagnosed 2007-2017)
- ○ SEER*Rx
- ○ SEER Manual
- ○ ICD-O-3 Update (for cases diagnosed 2018+)
- ○ Solid Tumor Rules (for cases diagnosed 2018+)

Staging

- ○ Collaborative Stage (for cases diagnosed 2016+)
- ○ Extent of Disease (EOD 2018)
- ○ Summary Stage 2018 (SS2018)

- ◉ Other

https://seer.cancer.gov/registrars/contact.html

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Hematopoietic and Lymphoid Neoplasms : Case Examples

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12/16/2020

Some guidelines on coding Hematopoietic Neoplasms

- Start with the Hematopoietic database
  - Find your working histology(ies)
  - Review information regarding
    - Primary Site
    - Transformations to
    - Transformations from
    - Same Primaries
    - Definition
    - Abstractor notes

Case 1: Question

- I have a Diffuse Large B-cell lymphoma (9680/3) DX 8/19/2011
- Then a low GR Follicular Lymphoma (9695/3) DX 4/17/18

- Multiple Primaries Calculator says the follicular is a new primary
- Morphology Code 1 9680/3 Diagnosis Year 1 2011
- Morphology Code 2 9695/3 Diagnosis Year 2 2018
- New Primary: I am confused by this because follicular transforms into Diffuse Large B-cell lymphoma. It confuse me that you would code this after because diffuse does not transform to follicular

Case 1: Answer

- This is a case of chronic (follicular) and acute (DLBCL), so rules M8-M13 would be the applicable rules. Since you have an acute followed by a chronic, Rules M12 and M13 apply to this situation
- If there was no treatment for the DLBCL, or you don't know, then Rule M12 applies and this is one primary, the DLBCL
- If there was treatment for the DLBCL, then Rule M13 applies and the follicular lymphoma is abstracted as a new primary

Case 2: Question

- Mediastinal lymphadenopathy, unilateral left sided pleural effusion, questionable left lung nodular abnormality, splenomegaly, thrombocytopenia, and nodes present above and below the diaphragm. Pericardial soft tissue nodularity
- Patient had biopsy of mediastinal nodes, which was positive for DLBCL
- What is the primary site

Case 2: Answer

- Rule PH21 applies: Primary site C778
  - Nodes present above and below the diaphragm
  - Mets: pleural effusion, pleural fluid, pericardial soft tissue nodule, left lung nodular abnormality
    - All these are recorded in Stage and do not factor into the primary site
  - Stage IV lymphoma

Case 3: Question

- Patient diagnosed with Primary Cutaneous Follicle Center Cell Lymphoma of the scalp bx 2/2020
- After PET shows diffuse lymph node involvement above and below diaphragm, axillary lymph node bx 5/2020 is positive for Follicular Lymphoma grade 2 AND 3 as well as DLBCL
- How many primaries do I abstract? I want to say per Rule M4 at least the Follicular / DLBCL is one primary, but is the Primary Cutaneous an additional primary based on Rule M10 and the time frame of 3 months between all of the diagnoses? That is where I am unclear….thank you for your insight

Case 3: Answer

- 2/2020: Primary Cutaneous Follicle Center Cell Lymphoma
- 5/2020 is positive for Follicular Lymphoma grade 2 AND 3 as well as DLBCL
- Start with the 5/20/20 diagnosis first
  - Per Rule M4, when there are two NHL's diagnosed simultaneously/same site-1 primary
  - Per Rule PH11, if one of the histologies is DLBCL, code to 9680/3

Case 3: Answer

- For 9597/3: Check to see if this histology transforms to DLBCL
  - No transformations listed for 9597
- Rule M15 applies: Use the multiple primaries calculator, comparing 9597/3 and 9680/3
  - Returns as new primary
- 1st primary: Primary cutaneous follicle center cell lymphoma (9597/3)
- 2nd primary: Diffuse large B-cell lymphoma (9680/3)

Case 4: Question/Answer

- If it stated **High grade B-cell lymphoma,** I would code 9680/3
- But it stated **Low grade B-Cell lymphoma or simply B-cell lymphoma,** I would code 9591/3
- The key to this is the “B-cell lymphoma”
  - A B-cell lymphoma stated as “low grade”, or with no other information (including a specific B-cell lymphoma) is coded to 9591/3
  - A B-cell lymphoma stated as “high grade” is an alternate name for 9680/3: Diffuse Large B-cell lymphoma
    - This terminology has become more common since the updated WHO Hematopoietic and Lymphoid Neoplasm book released in 2017
    - DLBCL may able be described as an “aggressive/very aggressive B-cell lymphoma”

Case 5: Question

- Oct 2015: C778 9680/3 DLBCL stage IIA treated with chemo and radiotherapy with complete response
- Dec 2017 PET shows involved LNs above and below diaphragm
- Jan 2018 Bx inguinal LN
  - -9680/3 DLBCL
  - -9698/3 Follicular Lymphoma, grade 3A
- Question: Is the Follicular Lymphoma considered a new primary based on rule M13?

Case 5: Answer

- First you must determine how many primaries are present in the second diagnosis before you can compare to the original diagnosis
  - Rule M4 applies, which states that if two or more non-Hodgkin lymphomas are present in the same biopsy, then you code one primary
- You are then referred to PH11 if one of the diagnoses is DLBCL. So, for the 2017 occurrence, the diagnosis is DLBCL
- Then you compare to the original diagnosis, which is DLBCL
  - Same primary per Rule M2 (same histology)

Case 6: Question

- I have in 2012 a Follicular lymphoma grade 1 (9695/3) with no further information, in 2018 the patient received chemo for a Diffuse large B-cell lymphoma (9680/3)
- In April 2020, the patient had a node biopsy who revealed a Follicular lymphoma grade 2 (9691/3), per rule M13 Abstract multiple primaries when a neoplasm is originally diagnosed as acute AND reverts to a chronic neoplasm after treatment
- With the mention of "originally", do I compare the last recurrence in 2018 or the first histology in 2012 with my new in 2020?

Case 6: Answer

- You need to compare the 2020 Follicular Lymphoma to both previous diagnoses
  - Rule M2 applies to the 2020 diagnosis, same histology as the 2012 diagnosis
- This is still two primaries
  - 2012: Follicular lymphoma
    - Reminder: The Follicular lymphomas are always the same primary (9690/3, 9691/3, 9695/3, 9698/3)
  - 2018: DLBCL

Case 7: Question/Answer

- What should be coded for Diagnostic Confirmation when a CLL/SLL patient is diagnosed by Flow Cytometry of peripheral blood only? FISH for peripheral blood with normal results and there was no tissue biopsy

- Recent confirmation (added to the Hematopoietic Manual)
  - We have recently received confirmation from our expert Hematopathologist on this very issue
  - If you have a peripheral blood smear and flow cytometry has been done on that, it would be a 3

Case 8: Question

- Year of Dx: 2019 Histology 9823/3
- Module 3, Rule PH5, Example states that positive peripheral smear with clinical LN involvement is primary site C421
- Rule PH6 indicates to code to involved LNs, organs, etc when no peripheral blood involvement AND no BM involvement or unknown if BM involvement
  - We are a bit confused by our case: Phys exam - 3cm submandibular mass - bx 1/10/19 shows Left submandibular LN bx: SLL/CLL
- Flow cytometry 1/30/19 shows immunophenotypic features consistent with CLL/SLL.
  CT C/A/P 1/30/19 - Mult normal size to mildly enlarged LNs compatible with lymphoma

Case 8: Answer

- CLL/SLL is frequently diagnosed via peripheral blood smear, and the flow cytometry is done on the peripheral blood smear
- So this case has positive peripheral blood involvement

- As a reminder, for CLL/SLL ONLY
  - Peripheral blood involvement or bone marrow involvement always take priority, even if you have microscopic confirmation of lymph node and/or organ involvement
  - For CLL/SLL: If peripheral blood or bone marrow is involved, primary site is C421. It does not matter what else is involved

Case 9: Question (A tough one)

- pt dx 2008 multi LNs w/ malignant lymphoma Non-Hodgkin, B cell (9591/3)
- 5/20/19 R axilla LN bx, - Diffuse large B-cell lymphoma arising in the setting of grade 3 follicular lymphoma
- The Hem data base states that Diffuse large B cell lymphoma(9680/3) can transform from Follicular lymphoma grade 3

- Since the path states Diffuse large B-cell lymphoma arising in the setting of grade 3 follicular lymphoma, isn't this then the same primary as the patient's diagnosis in 2008 , non-Hodgkin B cell lymphoma (9591/3)

- If it's not the same primary should the histology be coded as Follicular lymphoma grd 3(96983) since 9591/3 and 9680/3 are considered the same primary?

Case 9: Answer (A tough one)

- This is a tough case, because if you follow our rules strictly, you are going to have one primary; however, the fact that this patient first had an NHL (9591/3) and now has DLBCL (9680/3) shows that this is really a transformation
- Our rules are set up to account for about 80% of the cases, so it's not unheard for a case to not fit in nicely with the rules. As a side note, it is not possible to write rules that cover every possible scenario
- For your 2019 diagnosis, having a DLBCL arising in the setting of a follicular lymphoma would be one primary, the DLBCL
  - Rule M4 applies to this case, which is when two or more NHL's are present in the same anatomic structure at the same time, then you code one primary. PH11 states that if one of those histologies is DLBCL, then you code the DLBCL, which is the more aggressive histology

Case 9: Answer (A tough one)

- Having a diagnosis of a follicular lymphoma and a DLBCL diagnosed at the same time is becoming a more common occurrence based on the questions that we have been seeing
- Code the DLBCL (9680/3) as a second primary, which reflects the true clinical course that is happening with this patient. Although the database doesn't indicate that NHL, NOS (9591/3) transforms to DLBCL, it's important that the DLBCL information be captured because if this patient dies because of the lymphoma, it's going to be the DLBCL
- If you are able to follow back on the 2008 diagnosis and find a more specific lymphoma histology, that would be very helpful, although not absolutely necessary

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FABULOUS PRIZES

NAACCR

COMING UP!

- 3/4/21 Abstracting and Coding Boot Camp 2021
  - Patrick Stevens, CTR
  - Jim Hofferkamp, CTR
- 4/1/21 Larynx 2021
  - Denise Harrison, CTR
  - Louanne Currence, RHIT, CTR

CE'S

- Phrase
- Link

THANK YOU

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