

Hematopoietic and Lymphoid Neoplasms

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Outline

- Diagnostic confirmation
- New histologies (Effective for 1/1/2021+)
- Changes in reportable for existing histologies (Effective for 1/1/2021+)
- Primary Site and Histology coding examples
- Other Data Items and Hematopoietic Neoplasms

Diagnostic confirmation

Diagnostic Confirmation

- Hematopoietic manual: Coding Diagnostic Confirmation, Code 3 (pg. 19)
 - **New Note 1:** While every attempt is made to keep the Hematopoietic database updated, it is impossible to keep the Hematopoietic database updated with all the immunophenotyping or genetics that can be done for a specific histology since clinical medicine continues to evolve. If immunophenotyping or genetics are used by the pathologist/managing physician to identify a specific neoplasm that are not included in the Hematopoietic database, and Genetic testing and/or Immunophenotyping are listed as Definitive Diagnostic Methods for that histology, go ahead and use these

Diagnostic confirmation

- Review “Definitive Diagnostics Methods” section in the Hematopoietic Database
 - If genetics or immunophenotyping are listed, then the histology can have a diagnostic confirmation of 3
 - If genetics or immunophenotyping are not listed, then the histology cannot have a diagnostic confirmation of 3
 - If genetics or immunophenotyping are listed as definitive diagnostic methods, then review the “Genetics data” and “Immunophenotyping” fields in the database
 - If the pathology report lists one of these that confirms the diagnosis, then code 3 is applicable
 - If the pathology reports lists genetics or immunophenotyping that confirms the diagnosis and it is not listed in the Hematopoietic database, go ahead and assign 3
 - As noted in previous slide, it is very difficult to keep up with all the different tests used

Diagnostic confirmation

- New Note 2: These are histologies that are defined by genetics and must always have diagnostic confirmation code 3 (edits enforced for 2022+)
 - Two different notes in the Hematopoietic database
 - This histology can only be determined by positive genetics and/or immunophenotyping, diagnostic confirmation will always be 3.
 - Table B5: Myelodysplastic Syndromes (from Heme Manual): 9986/3
 - Primarily diagnosed based on bone marrow biopsy/peripheral blood
 - Table B7: Acute Leukemias of Ambiguous Lineage (from Heme Manual): 9806/3, 9807/3, 9808/3, 9809/3Pr
 - Primarily diagnosed based on bone marrow biopsy/peripheral blood
 - Table B8: Precursor Lymphoid Neoplasms (from Heme Manual): 9811/3, 9812/3, 9813/3, 9814/3, 9815/3, 9816/3, 9817/3, 9818/3, 9819/3
 - Primarily diagnosed on bone marrow biopsy/peripheral blood, but lymph node and/or organ tissue biopsies as well

Diagnostic confirmation

- For the histologies listed under Note 2 (edits enforced for 2022+)
 - These are histologies that are defined by genetics and must always have diagnostic confirmation code 3
 - Two different notes in the Hematopoietic database
 - This AML is part of the "AML with recurrent genetic abnormalities" group. Since this AML is diagnosed based on genetics, diagnostic confirmation will always be 3.
 - Table B6: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms : 9865/3, 9866/3, 9869/3, 9871/3, 9877/3, 9878/3, 9879/3, 9897/3, 9911/3, 9912/3, 9965/3, 9966/3, 9967/3
 - Primarily diagnosed based on bone marrow biopsy/peripheral blood

Diagnostic confirmation

- Examples of histologies that are always diagnostic confirmation 3:
 - 9986/3: Myelodysplastic syndrome with isolated del(5q)
 - This is MDS with a deletion on chromosome 5 (genetics)
 - 9807/3: Mixed phenotype acute leukemia with t(v;11q23); KMT2A-rearranged
 - This is an ambiguous leukemia with a KMT2A rearrangement (genetics)
 - 9814/3: B-lymphoblastic leukemia/lymphoma with t(12;21) (p13.2;q22.1); ETV6-RUNX1
 - This is a precursor lymphoid neoplasm with ETV6-RUNX1 and t(12;21)(p13;q22) (genetics)
 - 9866/3: Acute promyelocytic leukemias with PML-RARA
 - This is an AML that is part of the recurrent genetic abnormalities group

Diagnostic confirmation

- New Note 2: The following histologies should never be assigned diagnostic confirmation 3 since they are nonspecific codes and neither genetic testing or immunophenotyping are listed as Definitive Diagnostic Methods for these histologies. If there is immunophenotyping or genetics available, then a more specific histology code may be able to be assigned: 9590/3, 9655/3, 9800/3, 9820/3, 9860/3, 9863/3, 9980/3, 9982/3, 9989/3, 9991/3 (edits enforced for 2022+)
- This is a histology for which the Definitive Diagnostic Method does not include Genetics Data or Immunophenotyping, thus Diagnostic Confirmation should never be 3. If genetics and/or immunophenotyping are available, re-review to see if a more specific neoplasm can be coded.

Diagnostic Confirmation

- This note can be found in the following histologies:
 - 9590/3: Lymphoma, NOS
 - 9655/3: Hodgkin lymphoma, lymphocyte depletion, reticular
 - 9800/3: Leukemia, NOS
 - 9820/3: Lymphoid leukemia, NOS
 - 9863/3: Chronic myeloid leukemia, NOS
 - 9980/3: Myelodysplastic syndrome with single lineage dysplasia
 - 9991/3: Refractory anemia
 - (only used for cases diagnosed 2010-2020, code 9980/3 for 2021+)

Diagnostic Confirmation

- Remaining histologies (majority)
 - This histology can be determined by positive histology (including peripheral blood) with or without genetics and/or immunophenotyping. Review the Definitive Diagnostic Methods, Immunophenotyping and Genetics Data sections below, and the instructions in the Hematopoietic Manual for further guidance on assigning Diagnostic confirmation.
- For these histologies, genetics/immunophenotyping may be used that are not listed in the Heme DB
 - Per new Note 1, if this happens, that information can be used to code a 3
 - *Reminder:* Code 3 cannot be used when positive immunophenotyping or genetics testing that identified a more specific histology is preceded by ambiguous terminology
 - See Diagnostic Confirmation, “Assign code 3 for”, 1b.

Diagnostic Confirmation

- 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 [9823/3]) 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

Diagnostic Confirmation field in the Database (Histology 9865/3)

help me code for diagnosis year .

2021

Coding Manual: [Hematopoietic Coding Manual \(PDF\)](#)

Abstractor Notes

(This [code](#) is effective for [cases](#) diagnosed 2010 and later. For [cases](#) diagnosed prior to 2010, see [code](#) 9861/3.)

This AML has a generally poor [prognosis](#). Elevated [white blood](#) cell counts are most predictive of shorter overall [survival](#) and increased BM blasts are associated with shorter free [survival](#). Very limited [data](#) available regarding [treatment](#) or [survival](#).

In adults the [WBC](#) is generally lower than other AML types. The BM may have morphologic and cytochemical features of any FAB subtype of AML other than [acute](#) promyelocytic [leukemia](#) and [acute](#) megakaryoblastic [leukemia](#).

If the [leukemia](#) occurs before or [simultaneously](#) with Myeloid Sarcoma (9930/3), see M3 and Module 5:PH10.

Diagnostic Confirmation

This AML is part of the "AML with recurrent genetic abnormalities" group. Since this AML is diagnosed based on genetics, diagnostic confirmation will always be 3.

Grade

Not Applicable

Module 5:PH10



2021

Coding Manual: [Hematopoietic Coding Manual \(PDF\)](#)

Abstractor Notes

MDS with associated del(5q) is an MDS involves that [blood](#) and [bone marrow](#).

For MDS diseases (9980, 9982, 9983, [9985](#), 9986, 9989, 9991, 9992, 9993), [abstracting](#) each of the subtypes would result in over-counting of the diseases.

1. [Code](#) only the first subtype that is diagnosed.
2. Do not change the [histology code](#) or create a new [abstract](#) for any subsequent specific MDS subtypes

Recently the thalidomide [analog](#) lenalidomide has been shown to benefit MDS patients. [Transfusion](#) independence was achieved in two-thirds of patients.

Diagnostic Confirmation

This histology can only be determined by positive genetics and/or immunophenotyping, diagnostic confirmation will always be 3.

Grade

Not Applicable

Module Rule

None



Diagnostic Confirmation field in the Database (Histology 9590/3)

Coding Manual: [Hematopoietic Coding Manual \(PDF\)](#)

Abstractor Notes

This NOS [histology](#) is a [generic](#) disease description. [DCO cases](#) or path report only [cases](#) may stay in this classification. In most [cases](#), an NOS [histology](#) is only the working [diagnosis](#); the physician will run further [diagnostic procedures](#) and look for various clinical presentations to identify a more specific disease.

Further review of the [medical record](#) should be performed to look for the tests listed as [definitive diagnosis](#). When a more specific [diagnosis](#) is identified, the [histology](#) should be changed to the more specific neoplasm name and [code](#).

For [9590/3 malignant lymphoma](#), NOS, [non-Hodgkin lymphoma](#), classical [Hodgkin lymphoma](#), and any specific Hodgkin and non-Hodgkin lymphomas would be a more specific [histology](#).

See the [histology tables](#) ([Appendix B](#) of the [Hematopoietic manual](#)) for more information on NOS and more specific histologies.

Diagnostic Confirmation

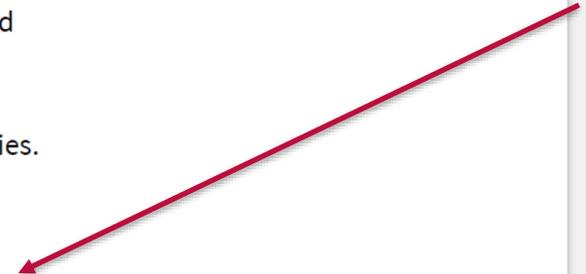
This is a histology for which the Definitive Diagnostic Method does not include Genetics Data or Immunophenotyping, thus Diagnostic Confirmation should never be 3. If genetics and/or immunophenotyping are available, re-review to see if a more specific neoplasm can be coded.

Grade

Not Applicable

Module Rule

None



Diagnostic Confirmation field in the Database (Histology 9680/3)

Coding Manual: [Hematopoietic Coding Manual \(PDF\)](#)

Abstractor Notes

Patients may present with nodal or **extranodal** disease. The most common **extranodal site** is the **gastrointestinal site** (stomach and ileocecal region). Other common **sites** of **extranodal** presentation include the **bone**, testes, **spleen**, **Waldeyer ring**, **salivary glands**, **thyroid**, **liver**, **kidneys**, and **adrenal glands**.

Patients usually present with a rapidly enlarging tumor **mass** at single or multiple nodal or **extranodal sites**. Many patients are **asymptomatic**, but **B symptoms** may be present. Specific localizing **symptoms** may be present and are highly dependent on the **site** of **extranodal** involvement.

For more information on **lymphoma**, see the **NCI** website: http://www.cancer.gov/types/lymphoma/hp/adult-nhl-treatment-pdq#section/_1 or http://www.cancer.gov/types/lymphoma/patient/adult-nhl-treatment-pdq#section/_129

Diagnostic Confirmation

This histology can be determined by positive histology (including peripheral blood) with or without genetics and/or immunophenotyping. Review the Definitive Diagnostic Methods, Immunophenotyping and Genetics Data sections below, and the instructions in the Hematopoietic Manual for further guidance on assigning Diagnostic confirmation.

Grade

Not Applicable

Module Rule

Module 6: PH11, PH13

Alternate Names



Changes in Histologies for 2021

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

- These are new, but they are not reportable to the standard setters
 - 9680/1: EBV-positive mucocutaneous ulcer
 - May *transform* to DLBCL (9680/3). The DLBCL would be the first reportable primary
 - 9738/1: HHV8-positive germinotopic lymphoproliferative disorder
 - Reportable form HHV8-positive diffuse large B-cell lymphoma (9738/3)
 - 9823/1: Monoclonal B-cell lymphocytosis, CLL-type
 - 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*
- Do not report these histologies. If your facility wants to report, do not report them as /2's, they are /1's, even with the "in situ" diagnosis
 - The /2 for in situ's applies to solid tumors only
- 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 Histologies (Effective 1/1/2021+)

- The following are new reportable histologies for 2021
 - 9715/3: Anaplastic large cell lymphoma, ALK-negative
 - For this diagnosis prior to 2021, code 9702/3
 - 9749/3: Erdheim-Chester disease (ECD)
 - Not reportable prior to 2021
 - 9766/3: Lymphomatoid granulomatosis, grade 3
 - For this diagnosis prior to 2021, code 9680/3
 - Note: Lymphoid granulomatosis, Grade 1 or 2, or NOS, are not reportable
 - 9819/3: B-lymphoblastic leukemia/lymphoma, BCR-ABL1-like
 - For this diagnosis prior to 2021, code 9811/3

New Histologies (Effective 1/1/2021+)

- For the following histologies, code 9861/3 for diagnosis prior to 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*
- 9968/3: Myeloid/lymphoid neoplasms with *PCM2-JAK2*
 - For this diagnosis prior to 2021, code 9975/3
- 9993/3: Myelodysplastic syndrome with ring sideroblasts and multilineage dysplasia
 - For this diagnosis prior to 2021, code 9985/3

Changes in Histologies (effective 2021)

- 9702/1: Indolent T-cell lymphoproliferative disorder of the gastrointestinal tract
 - Previously an alternate name for 9702/3 (2001-2020 in Heme DB)
- 9709/1: Primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder
 - Previously an alternate name for 9709/3 (2001-2020 in Heme DB)
- 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, except if diagnosed in Brain
 - *Note:* If PTLD is diagnosed with a lymphoma, still report the lymphoma (Rule M14)

Changes in Histologies (effective 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

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 important for
 - New histologies, obsolete histologies, changes in behavior

Help me code for diagnosis year :

2021



Case Examples

Primary Site and Histology Coding Examples

- When assigning primary sites:
 - It's important to know what are the common primary sites for the histology you are looking at
 - Primary site information available in the database
 - Note: This doesn't mean you can't have an uncommon primary site
- Instruction #4, Note 1 (pg. 35)
 - Do not simply code the site of a lymph node biopsy, use the information available from scans to determine the correct primary site
 - As a reminder, many times with lymphomas, they will biopsy the most convenient location to get a diagnosis
 - This does not mean this is the primary site
 - Always check your imaging

Preferred primary sites in Heme DB

Home Cancer Statistics ▾ SEER Data & Software ▾ Registry Operations ▾

[Search Database](#) [ICD-O-3 Code Lists](#)

Name
Classic Hodgkin lymphoma

ICD-O-1 Morphology Effective 1978 - 1991
9650/3: Hodgkin lymphoma, NOS

ICD-O-2 Morphology Effective 1992 - 2000
9650/3: Hodgkin lymphoma, NOS
9661/3: Hodgkin granuloma
9662/3: Hodgkin sarcoma

ICD-O-3 Morphology Effective 2001 and later
9650/3: Hodgkin lymphoma, NOS

Reportable
for cases diagnosed 1978 and later

Primary Site(s)
C770-C779

Lymph nodes (C770-C779) are the usual primary sites; however, involvement in others sites is possible. If you have confirmation that the only involved site is something other than the lymph nodes, then code to that primary site. See also Module 7.

For Hodgkin Lymphomas, the most common primary sites are the lymph nodes

This does not mean that Hodgkin lymphoma can't occur in other sites, just that it's very rare

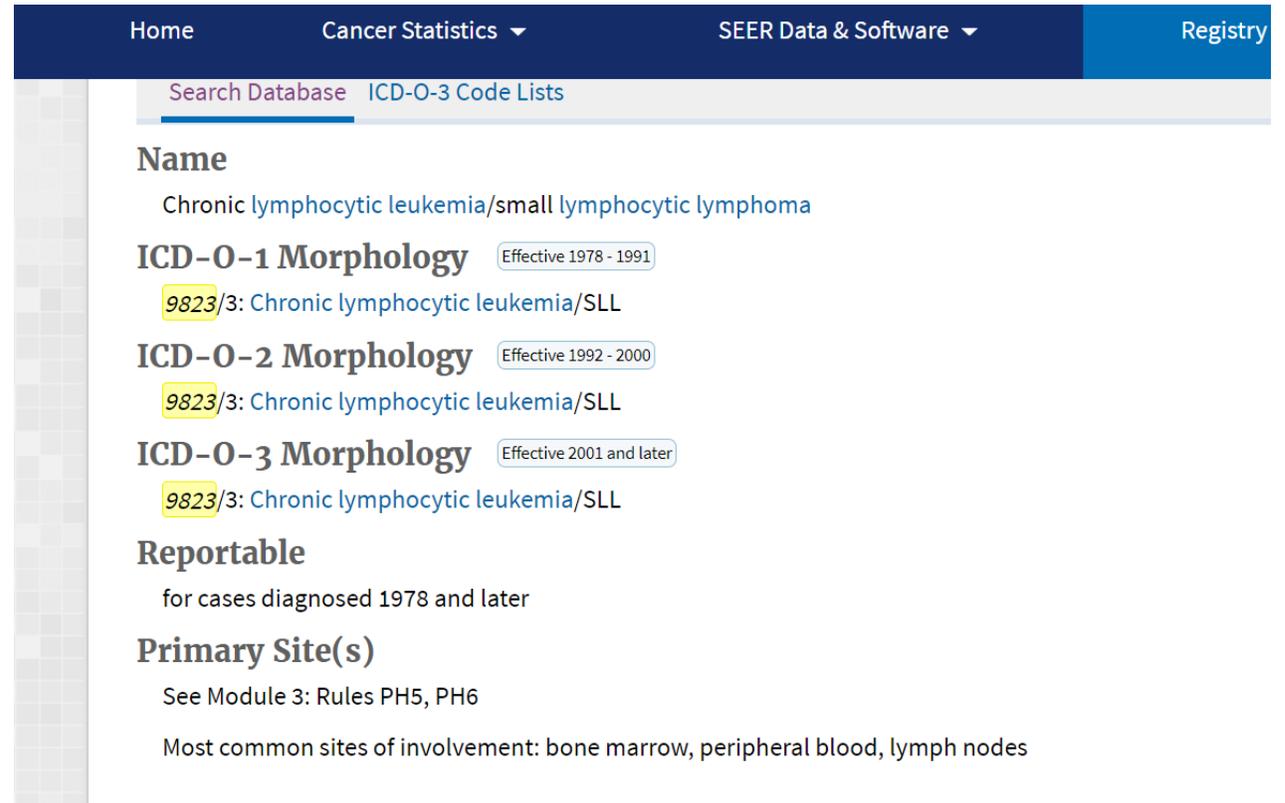
If lymph nodes are involved, along with other sites, then the other sites are going to be mets

Preferred primary sites in Heme DB

For CLL/SLL, the most common primary sites are the lymph nodes, bone marrow and peripheral blood

CLL/SLL can occur in other sites, but it's very rare

If one of the above is involved and there is additional involvement, then the other involvement is mets



The screenshot shows the SEER Data & Software Registry website. The navigation bar includes 'Home', 'Cancer Statistics', 'SEER Data & Software', and 'Registry'. The main content area is titled 'ICD-O-3 Code Lists' and displays the following information:

- Name:** Chronic lymphocytic leukemia/small lymphocytic lymphoma
- ICD-O-1 Morphology:** Effective 1978 - 1991. Code: 9823/3: Chronic lymphocytic leukemia/SLL
- ICD-O-2 Morphology:** Effective 1992 - 2000. Code: 9823/3: Chronic lymphocytic leukemia/SLL
- ICD-O-3 Morphology:** Effective 2001 and later. Code: 9823/3: Chronic lymphocytic leukemia/SLL
- Reportable:** for cases diagnosed 1978 and later
- Primary Site(s):** See Module 3: Rules PH5, PH6. Most common sites of involvement: bone marrow, peripheral blood, lymph nodes

Preferred primary sites in Heme DB

Home Cancer Statistics ▼ SEER Data & Software ▼

[Search Database](#) [ICD-O-3 Code Lists](#)

Name
Diffuse large B-cell lymphoma, NOS

ICD-O-1 Morphology
9612/3: Malignant lymphoma, immunoblastic type
9632/3: Malignant lymphoma, centroblastic type, NOS

ICD-O-2 Morphology Effective 1992 - 2000
9680/3: Malignant lymphoma, large B-cell, diffuse, NOS
9681/3: Malignant lymphoma, large cell, cleaved, diffuse
9682/3: Malignant lymphoma, large cell, noncleaved, diffuse
9712/3: Angioendotheliomatosis

ICD-O-3 Morphology Effective 2001 and later
9680/3: Malignant lymphoma, large B-cell, diffuse, NOS

Reportable
for cases diagnosed 1992 and later

Primary Site(s)
See Abstractor Notes and Module 7

DLBCL is a lymphoma that can occur anywhere in the body. It's one of the few lymphomas that can originate in the spleen, liver, brain, which are usually metastatic sites

Abstractor notes provide more information on the different types of presentations and the different terminology used for those

Primary Site and Histology Coding Examples

- Example: 2021 case I believe the primary site is skin RT thigh. The path report from the skin states consistent w/ NK T cell lymphoma. Bone marrow biopsy shows a NK T cell lymphoma . The imaging states infiltrative lesion soft tissues RT leg, inguinal , thoracic LN, lingula, RT tonsil, uvula.... The DR is calling this a cutaneous T cell lymphoma, NK/T cell origin
- The histology is 9719/3: Extranodal NK-/T-cell lymphoma
 - Per the Hematopoietic Database under “Primary Site(s)”
 - Most common sites of involvement: upper aerodigestive tract (nasal cavity, nasopharynx, paranasal sinuses, palate) with the nasal cavity (C300) being the prototypic site of involvement
 - Note: This scenario doesn’t fit the rules very well
 - Primary site is based primarily on the behavior of the histology

Primary Site and Histology Coding Examples

- Example cont:
 - Obviously for this histology, the skin (rt thigh) would not be the primary site, probably easiest accessible area of involvement to biopsy
 - Involvement of the skin would be recorded in staging and Mets at Dx fields
 - Based on information provided, imaging states soft tissue RT leg, inguinal, thoracic LN, lingula, RT tonsil, uvula
 - Head and Neck involved, which is part of the upper aerodigestive system
 - Since two parts of the Head and Neck are confirmed (by imaging) to be involved, best primary site would be C148: Overlapping sites of head and neck, pharynx
 - This primary site would be in line with the primary site information for this histology

Primary Site and Histology Coding Examples

- Example: Patient with brain tumor and mesenteric and retroperitoneal lymphadenopathy (which may be seen in the setting of lymphoproliferative process) on scan, surgery of the brain revealed High grade B cell lymphoma, physician not sure if this is CNS primary or stage IV. My question is the primary CNS primary with distant LN mets vs LNS (Mesenteric and Retroperitoneal) with CNS mets
- Physician doesn't even know if this is a primary lymph node primary or primary CNS
 - If the physician doesn't know, then we cannot presume and assign a primary site
 - For this case, the primary site would be unknown (C809) (Rule PH27)
- What if no statement from physician
 - Still go with C809 since High grade B-cell lymphoma is a histology that can originate in the brain (Rule PH27)
 - Note: many of the lymphomas are not going to originate in the brain, but 9680/3 (High grade B-cell lymphoma) does

Primary Site and Histology Coding Examples

- Patient diagnosed in 2020. She has a large mass in the vagina as well as mesentery with multiple lymph node regions involved including bilat cervical, rt atrium, lt atrium, rt pulmonary artery, rt internal mammary, rt mesenteric, bilat internal and external iliac and bilat inguinal.

I believe rule PH25 applies, and primary site would be coded to vagina although I am not sure. Please advise. The treating oncologist is staging as Stage IVE Burkitt's lymphoma of extranodal site.

The surgeon performing the biopsy states: Pelvic mass extending into the vagina which leads me to question should I code this to Pelvis nos rather than Vagina.

- Assign primary site as pelvis, NOS per the physician's statement
 - Vagina is not a common site for Burkitt, so this is extension into the vagina based on the physician's statement
 - Note: This scenario doesn't fit the rules very well, we are using the physician's statement

Primary Site and Histology Coding Examples

- 07/2020 Patient had a splenectomy w/ path positive for mantle cell lymphoma.
08/2020 Bone marrow biopsy positive for marrow involvement by mantle cell lymphoma. No imaging showing lymph node involvement. Physicians also do not mention that there was any involvement of lymph nodes. They just state spleen and bone marrow involvement.
- As a reminder, splenic involvement is usually metastatic disease
 - Note: There are two histologies that are specifically splenic lymphomas (noted in the database)
- For mantle cell lymphoma, a primary site of bone marrow is not common; however, it does happen (several questions received about this)
 - Code primary site to bone marrow (C421).
 - Splenic involvement would be metastatic disease
 - Note: This scenario doesn't fit the rules very well, although PH26 is close

Primary Site and Histology Coding Examples

- A.RIGHT LUNG (CORE BIOPSY): 6/9/2020: DIFFUSE LARGE B-CELL LYMPHOMA
PET: IMPRESSION:
 1. Hypermetabolic LAD in neck, chest, abdomen and pelvis, concerning for lymphoma.
 2. FDG-avid nodular soft tissue in RML, lingula, R posterior pleura, R anterior diaphragm, concerning for sites of extranodal lymphoma.
 3. Milder focus of uptake in L infraspinus may represent a site of IM lymphoma; physiologic or traumatic etiology is poss, but less likely given focal nature

- Primary site would be lung, based on Rule 25
 - Lung and its regional lymph nodes are involved
 - Distant lymph nodes and other involvement would be recorded in stage and Mets at Dx fields

Primary Site and Histology Coding Examples

- Patient with forehead/ scalp mass. Imaging shows LT Forehead mass INV SUBQ tissues above LT orbit EXT to calvarium W/O EXT to bone or intrathecal space. Also, EXT into superior medial LT orbit. MULT avid lesions LT parotid, LT parotid nodes, 12MM level 2A/B. Osseous lesion RT frontal calvarium. 8M RT axillary SFT tissue nodules. MULT avid osseous lesion BILAT femora and tibiae corresponds to SFT tissue marrow lesions on CT(Stated to have bone METS).
- Pathology states "integrated DX of AML t(9;11)(p21.3;q23.3); MLLT3-KMT2A Rearrange, manifesting as MYELOID SARCOMA, with unusually EXT monocytic DIFF/histiocytic transDIFF. The final histology is felt to represent Myeloid Sarcoma per MED ONC. A bone marrow biopsy is performed and is negative.
- Patient has AML with manifestations of Myeloid Sarcoma. This is one primary since the Myeloid Sarcoma is caused by the AML (See Rule PH10)
- Since you have a diagnosis of AML from the Soft Tissue, your histology is 9897/3: Acute myeloid leukemia with t(9;11)(p21.3;q23.3); KMT2A-MLLT3
 - Primary site is C421 (default for this histology), even though the bone marrow is negative

Primary Site and Histology Coding Examples

- Patient with history of Prostate cancer in 2002 treated with RT. In 2020 he had a bone marrow bx which showed: MDS with excess blasts-1 (MDS-EB-1). Immunostains show CD34-positive precursors are scattered and in small groups and are present at ~5% of cells. The findings are most consistent with a myelodysplastic syndrome, with excess blasts (MDS-EB-1). When patient was seen at my facility for consult the MD stated: Therapy related MDS with Del 3 and Del 7.

Is a physician statement enough to code treatment related MDS 9920/3, or should the code be 9983/3 MDS with excess blasts from the pathology report?

- Per Physician, this is therapy related MDS (C421 [default primary site], 9920/3)
 - Per Heme DB for 9920/3 (found under abstractor notes): If a specific myeloid neoplasm that is described with a different specific histology terms is also stated to be therapy related, code 9920/3 to capture the fact that this disease was therapy related. Document the other specific histology term in the text part of the abstract

Primary Site and Histology Coding Examples

- 05/03/219 Splenic mass, ultrasound guided fine needle aspiration biopsy, smears and sections: SMALL B-CELL NON-HODGKIN LYMPHOMA.
- 06/26/2019 Liver, mass, CT-guided needle biopsies: HEPATIC PARENCHYMA INVOLVED WITH THE PATIENT'S KNOWN SMALL B-CELL NON-HODGKIN LYMPHOMA, FAVOR SPLENIC MARGINAL ZONE TYPE.
- 07/30/2019 BM BX: NEG—documented Clin Hx—The patient was recently diagnosed with a small B-cell non-Hodgkin lymphoma in both liver and spleen biopsies The differential diagnosis included splenic marginal zone lymphoma and hairy cell leukemia. The current bone marrow biopsy shows a single small lymphoid aggregate in the biopsy without other evidence of lymphoma or lymphoproliferative disorder. Flow cytometry showed no B cell clonality
- 09/20/2019 Spleen: DIFFUSE LARGE B-CELL LYMPHOMA, GERMINAL CENTER TYPE. POSITIVE BY IMMUNOHISTOCHEMISTRY FOR CD20, CD79A, CD10, BCL2, BCL6, AND MUM1
- Do we accession this as a single DIFFUSE LARGE B-CELL LYMPHOMA, GERMINAL CENTER TYPE primary since they could not really classify the B-cell lymphoma until September?

Primary Site and Histology Coding Examples

- Example, cont.
- Per the pathology from May, there was a presence of a small cell lymphoma
 - DLBCL is not a small cell lymphoma, it is a large cell lymphoma. The DLBCL is transformation
- It is very rare for any lymphomas (other than the two splenic lymphomas) to originate in the spleen (although DLBCL can). It's also very rare for a lymphoma to originate in the liver. The fact that they confirm a small cell lymphoma in the spleen initially, then you are looking at a transformation from a small cell lymphoma to a large cell lymphoma
- Since there is evidence of lymphoma in the spleen and the physician states that they favor splenic marginal zone lymphoma, go ahead and assign the first primary to 9689 (primary site C422) and the second primary to 9680 (primary site C422). Splenic marginal zone lymphoma does transform to DLBCL

Other Data Items and Hematopoietic Neoplasms

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)
- The Mets at Dx fields for these histologies can be coded
 - No longer default to not applicable (edits updated)

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 (Bone, Brain, Liver, Lung, Distant Lymph Nodes, Other)

- Per the SEER 2022 manual ([SEER Program Coding and Staging Manual \(cancer.gov\)](https://seer.cancer.gov/manual/2022/))

- Use code 8 (Not applicable) for the following
 - i. Any case coded to primary site C420, C421, C423, C424, or C770-C779
 - ii. Plasma Cell Disorders 00822

- These instructions include the following:
 - All leukemias (primary site must be C421)



For questions about coding Hematopoietic Neoplasms (Primary Site, Histology, EOD/Summary Stage, Treatment)

Please post to Ask SEER Registrar

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

For Primary Site, Histology: Choose under “Reporting Guidelines”: Hematopoietic Rules (database and manual)

For EOD/Summary Stage: Choose under “Staging”, Extent of Disease or Summary Stage

For Treatment: Choose under “Reporting Guidelines”: SEER*Rx

Choose a subject

Please choose the most appropriate subject for your question. Hover over the **?** for subject if needed. **Questions submitted under the wrong subject require extra time to delayed response, as staff must manually triage your question.**

Reporting Guidelines

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

Staging

- Extent of Disease (EOD 2018)
 - Summary Stage 2018 (SS2018)
 - Collaborative Stage (for cases diagnosed 2016-2017)
- Other

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



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