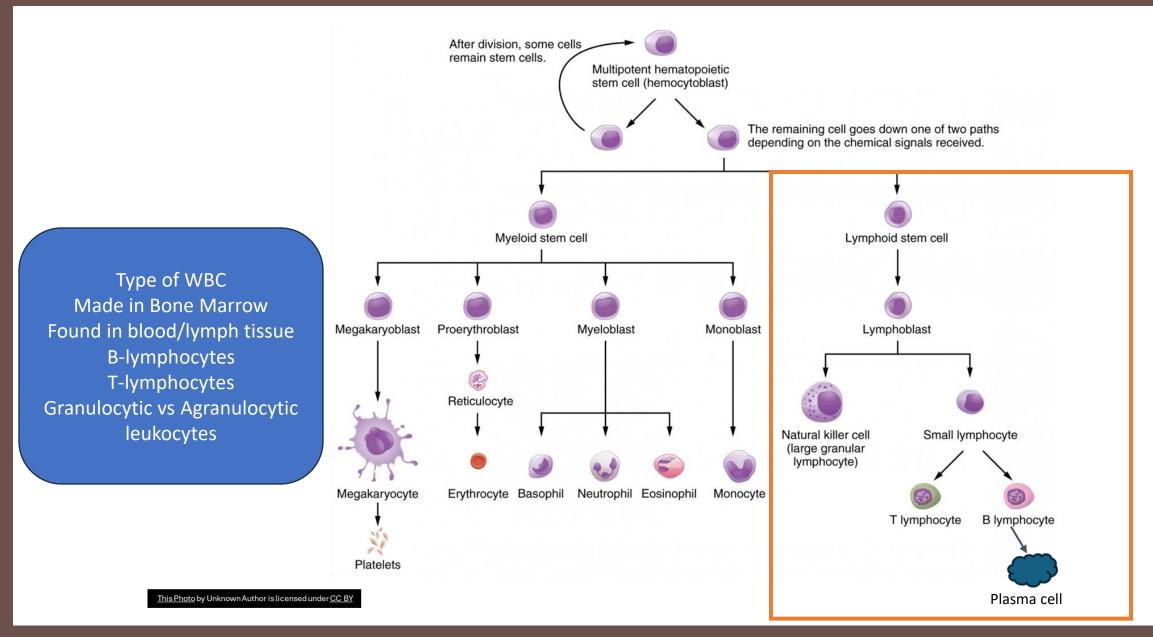


# Heme/Lymph: Lymphoid Neoplasms

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Presented by Melissa Riddle, ODS-C ICR Video Training Series | Iowa Cancer Registry March 2025



### Lymphoid Neoplasms

- Schemas v3.1 (2024+)
  - 00790 Lymphoma
  - 00795 Lymphoma-CLL/SLL
  - 00812 Primary Cutaneous Lymphoma
    - excl MF and SS
  - 00710 Lymphoma Ocular Adnexa
  - 00822 Plasma Cell Disorders
  - 00821 Plasma Cell Myeloma

# Lymphoma

Schema: 00790, 00795, 00812, 00710

# Introduction

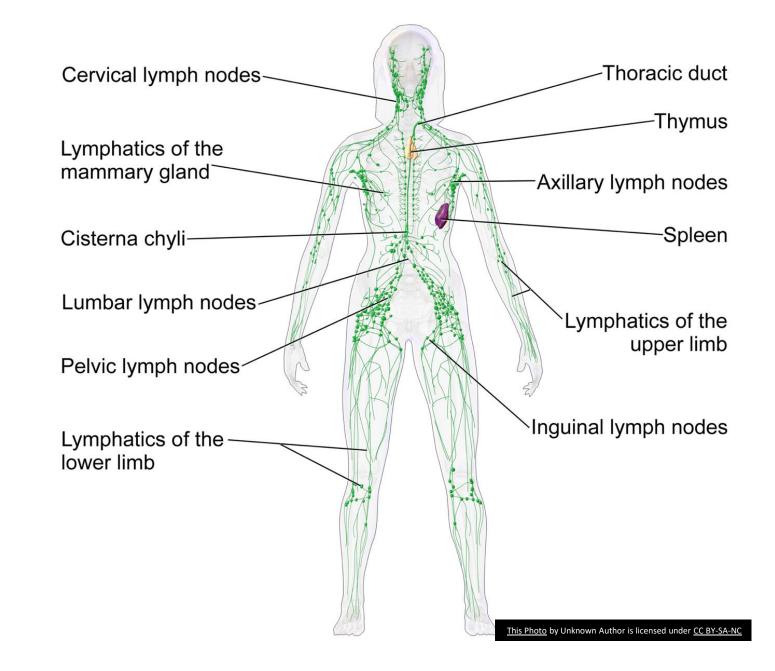
#### • What is Lymphoma?

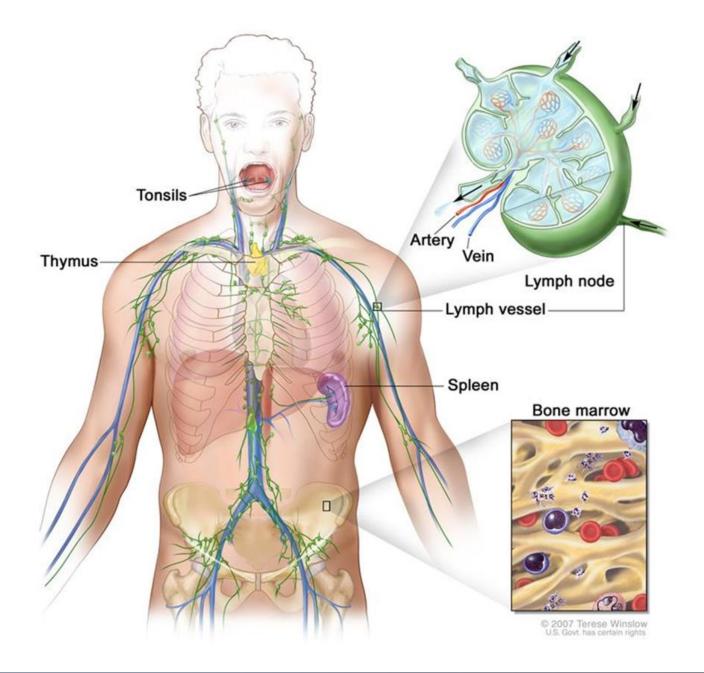
• A term that covers a variety of cancers of the lymphatic system.

#### • Two main groups:

- Hodgkin lymphoma (**HL**), AKA Hodgkin Disease
  - Originate almost anywhere in the body
- Non-Hodgkin lymphoma (NHL)
  - May originate in single lymph node, group of lymph nodes, or in another organ
  - Not a Hodgkin type

# Lymphatic System





#### Anatomy

The lymph system is made up of the following:

- Lymph: Colorless, watery fluid carries WBC's
- Lymph vessels: A network of thin tubes that collect and circulate excess fluid
- Lymph nodes: Small, bean-shaped structures that filter lymph and store white blood cells.
- Lymphoid organs: organs that are part of the lymphatic system such as thymus, bone marrow, tonsils, etc.
- Lymphatic tissue: part of the immune system

#### Lymphoid Tissue

Lymphoid tissue where **lymphocytes** and **plasma cells** are made. There are no regional lymph nodes in the brain, spinal cord, bone marrow, cartilage.

# Primary lymphoma occurs in:

#### • Lymph nodes

- Primary site is in the lymph node itself
- Part of lymphatic system

#### • Extranodal

- Sites outside lymph nodes
- Still part of lymphatic system.

#### • Extralymphatic

- Sites outside lymph system
- Common sites include GI tract, lung, thyroid, brain.

#### Lymphatic Organs & & Extranodal Sites

**Spleen**: left side of the abdomen, filters blood. Largest lymphatic organ.

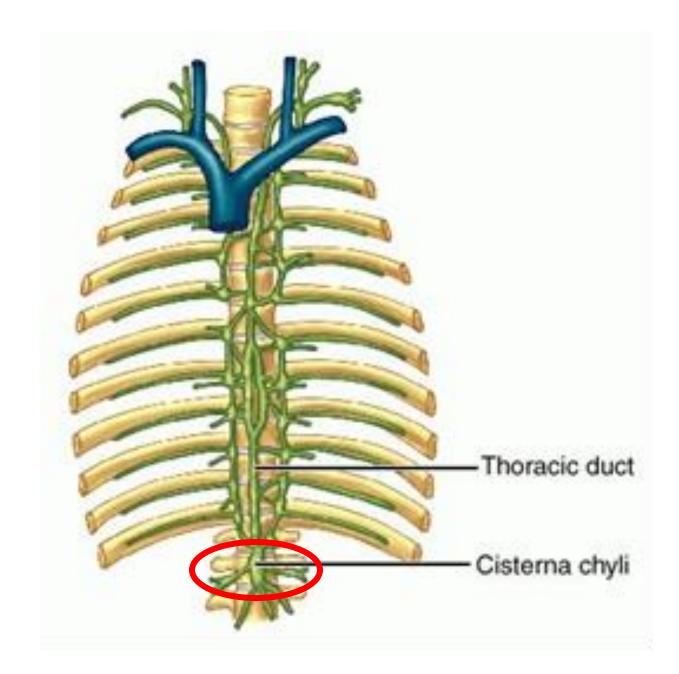
**Thymus**: in the chest behind the breastbone, processes T-lymphocytes, produces hormone.

Tonsils: lymph tissue at the back of the throat

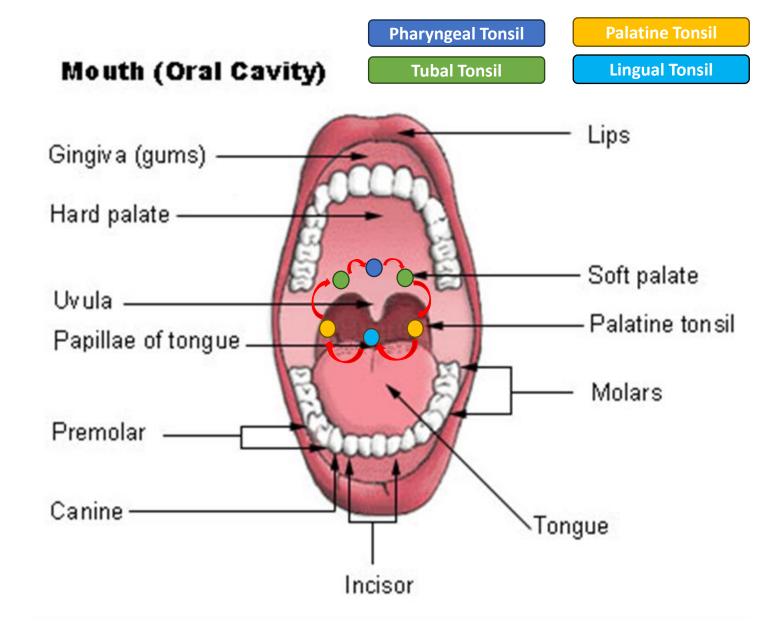
**Peyer's Patch**: visceral surface of the small intestine, usually ileum.

All part of the lymphatic system

### Anatomy



#### Waldeyer's Ring



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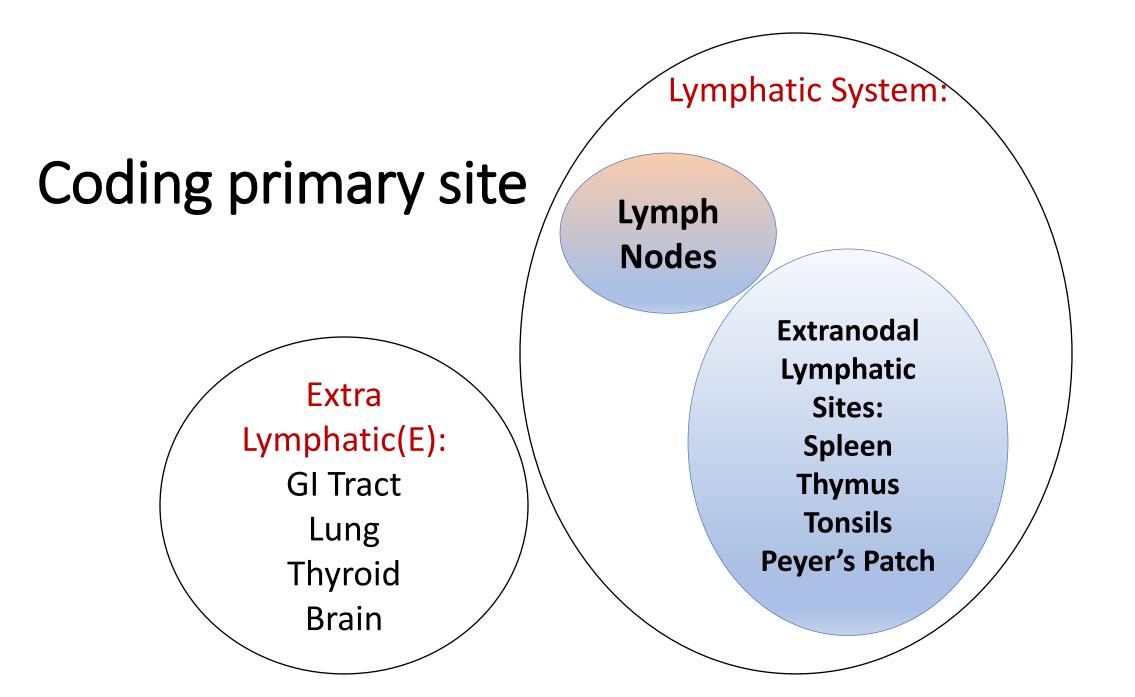
han Secretory IgA San Antigen Mucus M cell Epithelial cell T cell Plasma cell B cel Peyer's patch Organized lymphoid follicles > Lymphatic system T cell B cell - MHC II B cell receptor Dendritic cell

### Peyer's Patch

# Site & Morphology

Assistance for determining codes for:

- LN Chains
  - Use Appendix C | Heme Manual
- Primary Site/Histology
  - Use Heme-Lymph Database and Manual



#### Lymph node Topography Codes

Code	Site
C77.0	Lymph nodes of head, face and neck
C77.1	Intra-thoracic lymph nodes
C77.2	Intra-abdominal lymph nodes
C77.3	Lymph nodes of axilla or arm
C77.4	Lymph nodes of inguinal region or leg
C77.5	Pelvic lymph nodes
C77.8	Lymph nodes of multiple regions
C77.9	Lymph node, NOS

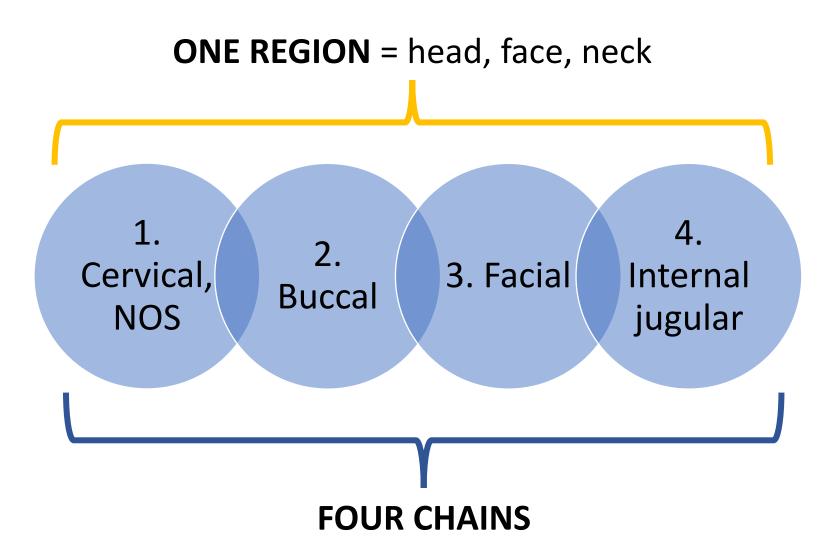
#### **Primary Site**

- Per Jennifer Ruhl: Not all sites of involvement are used to assign primary sites. Some sites of disease may be metastatic.
- Module 7 (PH18-PH27)
- *Note 2*: Do not simply code the site of a biopsy; use the information available from imaging to determine the correct primary site
- Note 3: Secondary involvement of distant lymph nodes (for an extranodal lymphoma), bone marrow, liver, spleen or CNS are included in the stage fields only. This secondary involvement excludes rare primary lymphoid neoplasms of spleen, multifocal 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.

## **Primary Site**

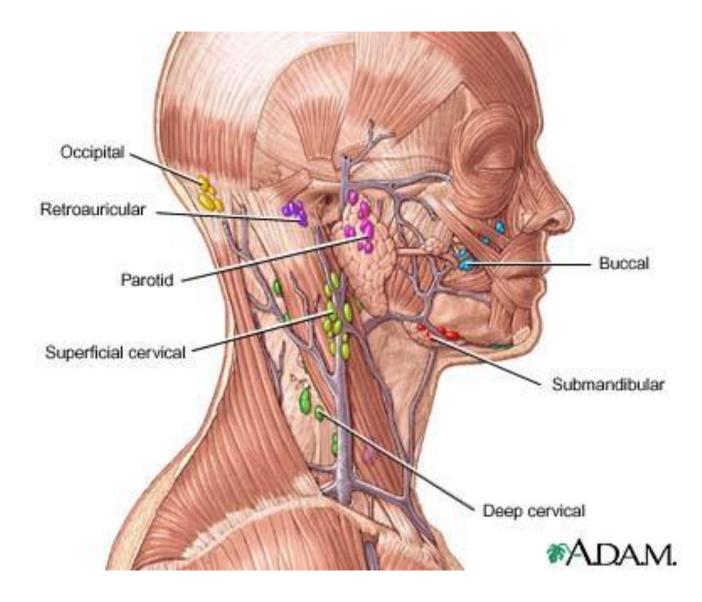
- Use imaging to determine your primary site
  - Also used to determine stage
  - This is critical for lymphomas
- Site of biopsy is confirming the diagnosis, it is not necessarily your primary site
  - These are very different from solid tumors
  - Biopsy is often the most convenient location to assess
  - **<u>ALWAYS</u>** review your imaging

### Chains vs Regions



# **Primary Site**

All the same "region", code to c770

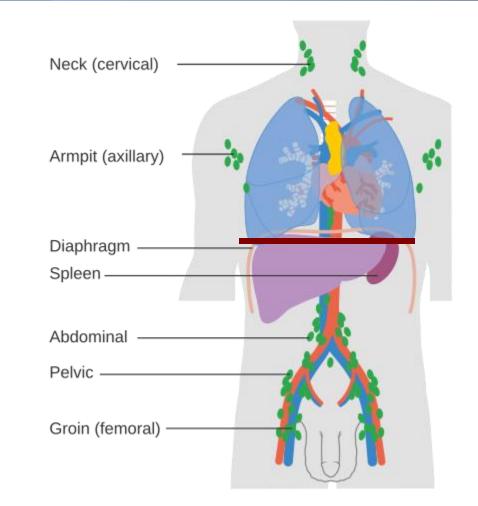


#### Heme/Lymph Manual – Appendix C

Lymph Node/LN Chain	Site	ICD-O LN Region(s)	TNM Staging
Anterior deep cervical	C770	Head, face, neck	Cervical, right & left*
Anterior jugular	C770	Head, face, neck	Cervical, right & left*
Buccal	C770	Head, face, neck	Cervical, righ & left*
Central Compartment	C770	Head, face, neck	Cervical, right & left*
Delphian node	C770	Head, face, neck	Cervical, right & left*
Esophageal groove	C770	Head, face, neck	Cervical, right & left*

### Case Example - Lymphoma

- Workup: supraclavicular necrotic mass
  - LAD above and below diaphragm suspicious for malignancy
  - Biopsy of supraclavicular LN follicular lymphoma
  - No statement of where this originated
- Step 1: Provisional histology
  - Follicular lymphoma 9690
- Step 2: MP Rules
  - Single primary **M2**
- Step 3: PH Rules
  - Module 7 PH21 code primary site to multiple LN regions C778
    - Multiple LN regions involved (defined by ICD-O)
    - Not possible to identify LN region where originated



### Non-Hodgkin Lymphoma (NHL)

#### • Histology 9591 (NHL, NOS)

- Schema Discriminator 1 necessary for accurate Stage Group table use
  - Splenic B-cell lymphoma/leukemia unclassifiable = abstract/stage as leukemia
  - Hairy cell leukemia variant = abstract/stage as leukemia
  - Splenic diffuse red pulp small B-cell lymphoma = abstract/stage as lymphoma
  - Non-Hodgkin Lymphoma NOS = abstract/stage as lymphoma

Code	Description	Schema ID #/Description
1	Splenic B-cell lymphoma/leukemia, unclassifiable	00830: HemeRetic
2	Hairy cell leukemia variant Prolymphocytic variant of hairy cell leukemia	00830: HemeRetic
3	Splenic diffuse red pulp small B-cell lymphoma Splenic marginal zone lymphoma, diffuse variant Splenic red pulp lymphoma with numerous basophilic villous lymphocytes Splenic lymphoma with villous lymphocytes	00790: Lymphoma (excluding CLL/SLL)
9	Non-Hodgkin lymphoma, NOS Any other terminology describing non-Hodgkin lymphoma, NOS	00790: Lymphoma (excluding CLL/SLL)
<blank></blank>	Histology is NOT 9591, Discriminator is not necessary	



# Staging

Lymphoma Schema 00790

### Lymph Node Involvement

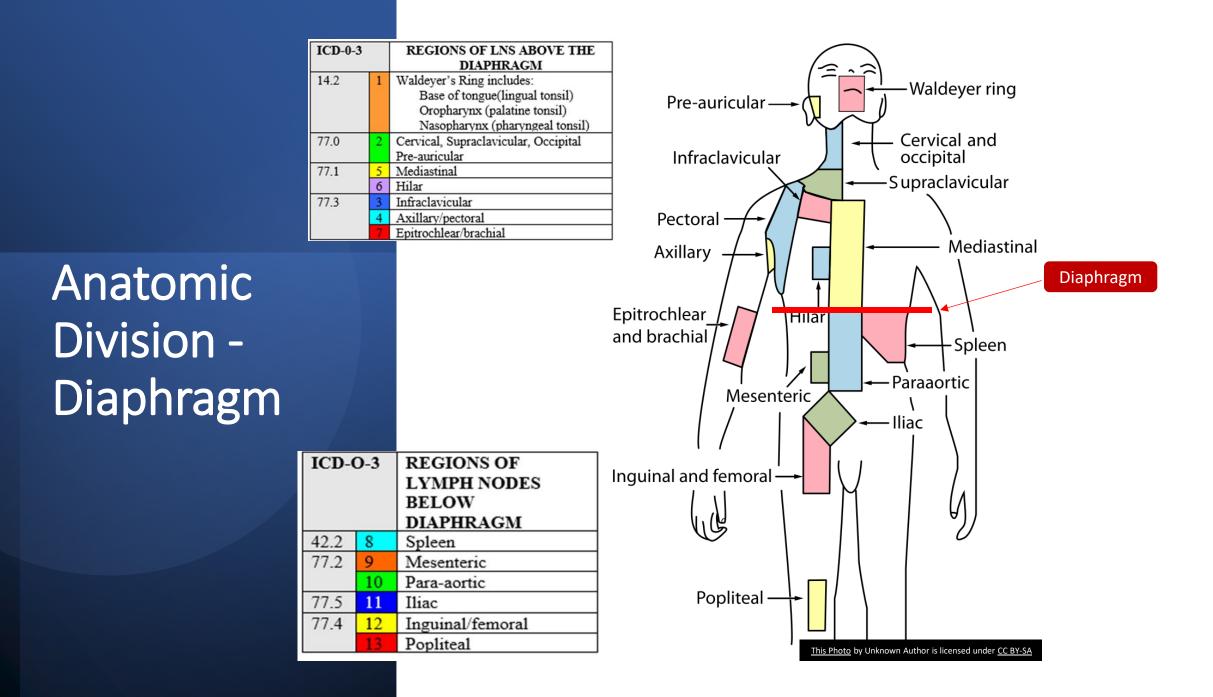
#### SEER\*RSA v3.1

#### **EOD Primary Tumor Lymphoma schema**

 Note 4: Any mention of the terms including fixed, matted, mass in the hilum, mediastinum, retroperitoneum, and/or mesentery, palpable, enlarged, shotty, lymphadenopathy are all regarded as involvement for lymphomas when determining appropriate code.

# **Bulky Disease**

- EOD Primary Tumor
  - Note 5: "Bulky Disease" (500)
    - Varies by lymphoma histology
      - <u>Hodgkin</u>: the ratio between the maximum diameter of the mediastinal mass and maximal intrathoracic diameter based on CT imaging in Lugano classification
      - <u>Non-Hodgkin</u>: main criteria is based on size with cutoffs ranging from 5-10cm, although 10cm is recommended
    - Typically stated by physician or on imaging study



### Nodal Lymphoma Staging

#### Axial Lymph Nodes

- Staging: Each region counts a one LN region
  - LN Region examples: Mediastinal, Hilar, Para-aortic, Retroperitoneal, Mesenteric, Internal mammary

#### **Bilateral Lymph Nodes**

- Staging: If LNs on both sides (bilateral) are involved count these as 2 regions
  - LN Region examples: Cervical, Supraclavicular, Occipital, Preauricular, Infraclavicular, Axillary, Inguinal/Femoral

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

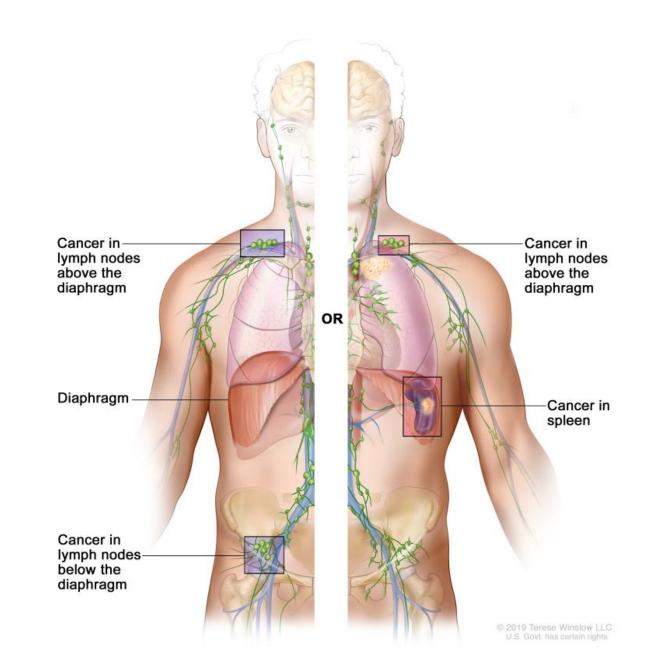
\*The right and left are separate regions per AJCC

Lymph Node/Lymph Node Chain	Use for Multiple Primaries in Heme	ICD-O Lymph Node Region(s)	TNM Staging
Abdominal	C772	Intra-abdominal	Mesenteric
Anorectal (pararectal)	C775	Pelvic	Pelvic, right and left*
Anterior axillary (pectoral)	C773	Axilla or arm	Axillary, right and left*
a . a . a <i>b d</i> . as			

- Appendix C
  - Bilateral
    - Left and right axillary LN = 2 REGIONS for staging EOD & AJCC

# LN Chains – Appendix C

Regions separated by right and left <u>AND</u> Regions separated by diaphragm



Code	Description	SS2018 T
100	Nodal lymphomas Single lymph node region involved	L
	> Involvement of multiple nodal chains in the SAME lymph node region	
200	Extranodal lymphomas   Single extralymphatic site  WITHOUT nodal involvement  Multifocal involvement (except multifocal lung involvement or any liver involvement, see code 800) of one extralymphatic organ/site WITHOUT nodal involvement (see code 400 for WITH nodal involvement)	L
300	Nodal lymphomas   Two or more lymph node regions involved  SAME side of diaphragm	RE

Bilateral Cervical Lymph node involvement

• EOD 300 [bilateral makes this 2 regions]

- Code 800 for the following:
  - Liver enlargement is not enough alone to code as involvement
    - Involvement is indicated by diffuse uptake or mass lesion or abnormal liver function tests; liver biopsy confirm involvement
      - Includes liver primaries of lymphoma
  - Lung involvement is indicated by pulmonary nodules or parenchymal involvement on PET or CT in absence of other causes
    - Lung biopsy may confirm
      - May be <u>code 700</u> (not multifocal)
      - *Mets at DX Lung* = 1
  - Bone involvement (NOT including bone marrow) indicated by avid lesion on PET
    - Bone biopsy may be used to confirm equivocal involvement
      - *Mets at DX Bone* = 1
  - CNS involvement is often suspected due to symptoms and can be confirmed via imaging
    - CSF by flow cytometry may be done
      - *Mets at DX Other* = 1

#### • Peripheral Blood involvement

- Assessed by aspiration or peripheral blood smear
- Primary site C421
  - Do NOT code *Mets at DX-Other* as 1
- Peripheral blood smear not done but physician clinical assessment indicates involvement this can be used
- If **ONLY** the peripheral blood is involved **Code 750**
- Peripheral blood involvement <u>WITH</u> other involvement Code 800

#### Bone Marrow involvement

- Primary site is NOT C421 Code 800
  - *Mets at Dx Other* = 1
- No bone marrow biopsy/aspiration performed but physician clinical assessment indicates involvement, code as involved
- If only bone marrow involved, code primary site to C421, EOD PT: 800
  - Do NOT code *Mets at Dx Other* as 1

Code	Description	SS2018
100	Nodal: Single LN region involved; involved multiple nodal chains in SAME region	L
200	Extranodal: Single extralymphatic site w/o nodal involvement Multifocal involvement of one extralymphatic organ/site w/o nodal involve (see <i>code 400</i> with nodal involvement)	L
300	Nodal: Two or more LN regions involved, SAME side diaphragm	RE
400	<ul> <li>Nodal: Contiguous extralymphatic extension from nodal/lymphatic site</li> <li>With or without involvement of other nodal regions; SAME side diaphragm</li> <li>Extranodal:</li> <li>Localized involvement of single extralymphatic organ/site</li> <li>WITH involvement of its regional LN(s) OR</li> <li>WITH involvement of other LN(s) on SAME side of diaphragm</li> </ul>	RE
500	Code 300 or 400 with <b>bulky disease</b>	RE

Code	Description	SS2018
575	<ul> <li>Nodal and Extranodal lymphomas</li> <li>Involvement of LN regions BOTH sides diaphragm</li> <li>Without or unknown spleen</li> </ul>	D
600	<ul> <li>Nodal: Involvement of LN regions on BOTH sides of diaphragm</li> <li>OR nodes ABOVE the diaphragm involved</li> <li>WITH spleen involvement</li> </ul>	D
700	<ul> <li>Diffuse or disseminated (multifocal) involvement of ONE extralymphatic organ/site</li> <li>WITH or WITHOUT assoc LN involvement</li> <li>See other notes</li> </ul>	D
750	Peripheral blood involvement <u>ONLY</u>	D
800	Diffuse or disseminated (multifocal involv) of Bone, CNS. Any involvement of bone marrow, CSF, liver, lung (multiple lesions other than 400), peripheral blood involvement WITH other involvement. Distant Mets NOS	D
999	Unknown, Not documented, cannot be assessed	D

#### EOD Reg LN and Mets

Code	Description	SS2018
888 (Reg LN) 88 (Mets)	Not applicable: Info not collected for this schema	NA

### Lymphoma - SSDI

#### **B** Symptoms

#### **HIV Status**

NCCN International Prognostic Index (IPI)

Code	Description
0	No B symptoms (asymptomatic); classified as A by physician when asymptomatic.
1	Any B symptom(s) Night sweats (drenching); Unexplained fever or wt loss, B symptoms NOS. Classified as B by physician when symptomatic.
9	Not documented in med rec. B symptoms not assessed or unknown if assessed. No mention of B symptoms.

*Note 1*: Physician statement of B symptoms can be used when no other info available.

*Note 2*: Each stage classified as A or B according to absence or presence of defined constitutional symptoms: fevers, night sweats, wt loss.

*Note 3*: Pruritus alone not a B symptom.

Note 4: Code 9 if no mention of B symptoms.

#### SSDI: B symptoms

Code	Description	
0	Not assoc with HIV/AIDS; HIV neg	
1	Associated with HIV/AIDS; HIV pos	SEER requires data items 2018+
7	Test ordered, results not in chart	CoC required 2018-2020
9	Not documented in record HIV status not assessed or unknown if assessed	

*Note 1*: Physician statement of HIV status can be used if no other info.

*Note 2*: AIDS lymphomas are late manifestation of HIV infection and have unique and clinical and path features that differ from lymphoma in general population.

*Note 3*: HIV includes types I and II. Older term include HTLV-3 and LAV.

Note 4: Code 9 if no mention of HIV/AIDS in med record. Do not assume pt is HIV neg.

*Note 5*: Hx of HIV, assign <u>code 1</u>, even if not currently detectable.

#### **SSDI: HIV Status**

Code	Description	
00-88	0-8 points	
X1	Stated low risk (0-1 points)	For NHL histologic types, not for
X2	Stated as low intermediate risk (2-3 points)	Hodgkin's
X3	Stated as intermediate risk (4-5 points)	Lymphoma
X4	Stated as high risk (6-8 points)	
X9	Not documented in record, not assessed, unknown if assessed;	Hodgkin Lymphoma

Note 1: <u>Physician statement of NCCN IPI must be used to code this item</u>. Do not calculate points or assign risk. Only record points or risk if physician has documented them. Use points over risk if both are available.
Note 2: NCCN IPI is applicable for NHL only. If you have score for HD (IPS) <u>do not code that info here</u>
Note 3: Low, intermediate, high risk assoc with RAI Stage not recorded here.

### SSDI: IPI

# Treatment

NCCN Guidelines

#### Treatment: Adult NHL

#### Depends on stage

- Radiation Therapy
- Chemotherapy
- Targeted therapy (monoclonal antibody)
- Watchful waiting

New types of treatment in Clinical trials:

Vaccine therapy (biological), high-dose chemo with stem cell transplant

### Surgery– Nodal Lymphoma

#### • Surgery of Primary Site:

• LN Surg Codes C770-C779

#### Scope of REG Lymph Node surgery

- Code 9 for schemas:
  - 00790 Lymphoma (C770-C779 only; excl CLL/SLL)
  - 00795 Lymphoma CLL/SLL

#### • Surgical Procedure of Other Site:

• Standard Codes

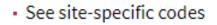
#### Lymphoma

Coding Guidelines: Lymphoma (PDF, 182 KB)

Hematopoietic and Lymphoid Neoplasm Coding Manual (PDF, 1.0 MB)

#### Surgery Codes

• Lymph Nodes - (C770-C779) (PDF, 148 KB)



#### SEER Appendix C

### Nodal Lymphoma Surgery Codes

Description	If mult LN	Notes	Code
Local tumor excision NOS	involved and only one removed – code as biopsy	Less than a full chain <b>Primary in one and <u>ONLY</u> one LN</b> – this node <u>removed</u> by excisional bx	A250
<ul><li>LN Dissection NOS</li><li>One chain</li><li>Two or more chains</li></ul>			A300 • A310 • A320
<ul><li>LND + Splenectomy</li><li>One chain</li><li>Two or more chains</li></ul>			A400 • A410 • A420
<ul> <li>LND <u>and</u> Partial/Total removal adjacent organ(s)</li> <li>One chain</li> <li>Two or more chains</li> </ul>			A500 • A510 • A520
<ul> <li>LND <u>and</u> Partial/Total removal adjacent organ(s)</li> <li>One chain</li> <li>Two or more chains</li> </ul>	+ Splenectomy	Includes staging laparotomy for lymphoma	A600 • A610 • A620

### Nodal Lymphoma

Scope of Regional LN Surgery:	<b>Code 9</b> – C770-C779
Regional LN Pos/Exam:	<b>99/99</b> – C770-C779 • Lymphoma 00790 • Lymphoma CLL/SLL 00795
Surgery Other Site:	Standard codes

#### Splenic Lymphoma Surgery Codes

#### SEER Appendix C: Other & Unknown Sites Spleen Surgery Codes:

Description	Code
Partial splenectomy	A210
Total splenectomy	A220
Splenectomy NOS	A800

Scope Regional LN Surgery: Standard codes Surgery of Other Site: Standard codes

#### Surgery Coded Fields – Extranodal or Extralymphatic lymphomas

#### • Surgery Primary Site:

 Use surgery codes for specific <u>Primary Site Identified</u> (i.e. stomach lymphoma, use gastric surgery codes)

#### • Scope of Reg LN:

- Standard Codes
- Surgery of Other Site:
  - Standard Codes

### NHL – NCCN Guidelines

#### • Follicular Lymphoma

- <u>Slow-growing</u>
  - Stage I or II possibly no treatment (watch and wait)
    - Radiation
    - Rituxan with or without chemo
- Fast-growing
  - Stage III or IV often treated more like DLBCL
    - Watch and wait
    - Chemo and/or radiation

#### • DLBCL

• Per stage, R-CHOP common

Treatment: Hodgkin Disease

- Chemotherapy/Hormone
- Radiation therapy
- Surgery

#### New treatments being tested:

- Chemo and RT with stem cell transplant
- Monoclonal antibody therapy

### Hodgkin Lymphoma

#### • NCCN Guidelines

- Depends on Stage
- Chemo [multi-drug regimen] with or without Prednisone
- Radiation Therapy
  - [ISRT] Involved Site Radiation Therapy recommended to treat HL
  - Uses EBRT & MRI for enhanced planning techniques
- Stem cell Transplant

## Lymphoma Cases



#### Lymphoma Case 1

3/10/2024 32yo with enlarged cervical lymph node with complaint of fatigue, drenching night sweats, and intermittent fevers for 3 mo; PE: palpable enlg R cervical node, 1cm; small palpable bilateral axillary LN; Plan: biopsy cervical node and imaging

3/16/2024 cervical node bx: diffuse large B-cell lymphoma

3/24/2024 CT C/A/P: multiple enlarged right cervical nodes, bilateral axillary lymphadenopathy as well as slight enlarged retroperitoneal LN

3/26/2024 PET: multiple involved lymph nodes in the right cervical, bilateral axilla, retroperitoneum, and mediastinum suspicious for lymphoma

4/18/24 bone marrow bx: follicular lymphoma

#### Lymphoma Case 1 cont.

3/31/24 Med Onc Note: Newly diagnosed patient with DLBCL involving multiple lymph nodes; Plan: bone marrow biopsy and determine treatment

4/28/24 Med Onc Note: DLBCL patient with involvement of multiple lymph node regions and bone marrow; IPI score 5; Plan: R-CHOP

5/5/24 Start R-CHOP

#### Lymphoma Case 1

Data Items	Primary 1 Codes	Primary 2 Codes
Primary Site		
Histology		
EOD Primary Tumor		
B-Symptoms		
HIV Status		
IPI		
Chemotherapy		
Hormone Therapy		
Immunotherapy		

#### Lymphoma Case 2

2014 – NHL (9591/3) C778 in the registry 2024 - CD30+ lymphoproliferative disorder

Is the 2024 case a new primary? MP Rule: Primary Site: Histology:

#### Lymphoma Case 3

2020 – T-cell/histiocyte rich large B cell lymphoma, C771 (in registry)
5/2024 Bone marrow bx: DLBCL
5/2024 CT C/A/P: no lymphadenopathy noted and no other abnormalities

Is the 2024 case new?

MP Rule:

Primary Site 2024:

Histology 2024:

# Lymphoma CLL/SLL

Schema ID 00795

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### CLL/SLL

- Chronic lymphocytic leukemia/small lymphocytic lymphoma (CLL/SLL)
  - Can be this combined histology or
  - Either of these
  - Histology: 9823/3
- Frequently diagnosed by flow cytometry (immunophenotyping)
  - Bone marrow biopsy or other biopsy
- Module 3: PH5, PH6

#### **General Notes**

- SEER\*RSA v3.1 Lymphoma CLL/SLL
  - *Note 3*: CLL/SLL is always staged as a lymphoma.
  - *Note 5*: In addition to coding EOD Primary Tumor, the following are needed to assign a stage group for CLL/SLL:
    - $\circ$  Lymphocytosis
    - $\circ$  Adenopathy
    - $\circ$  Anemia
    - Organomegaly
    - $\circ$  Thrombocytopenia

### SSDI for Lymphoma CLL/SLL

B symptoms	
HIV Status [COC 2018-2020 only]	Same as lymphoma
NCCN IPI	SSDIs
Lymphocytosis	
Adenopathy	
Organomegaly	
Anemia	
Thrombocytopenia	

#### Lymphocytosis

**Note 1**: All cases CLL/SLL require Lugano classification, captured in AJCC stage group and 5 components of modified RAI staging system.

*Note 2*: Rai stage only for CLL with C421. See code 5.

*Note 3*: Lymphocytosis (lymphocyte number) is defined by an absolute lymphocyte count (ALC) >5,000.

*Note 4*: Record this item based on CBC and diff performed at dx and pre-rx. In absence of lab test, use phys statement.

*Note 5*: No mention of lymphocytosis, code 9.

*Note 6*: Physician stated Rai takes priority when conflicting info.

Code	Description
0	Lymphocytosis not present; ALC <= 5,000
1	Lymphocytosis present; ALC>5,000
5	Not applicable: Primary site not C421
6	Lab value unkn, phy states lymphocytosis present Physician stated Rai stage 0-IV
7	Test ordered, results not in chart
9	Not documented in record. Lymphocytosis not assessed, unknown if assessed. [no mention]

### Adenopathy

*Note 1*: All cases **CLL/SLL require Lugano classification**, captured in AJCC stage group and <u>5 components of modified RAI staging</u> <u>system</u>.

*Note 2*: Rai only applicable for CLL, site C421. See code 5.

*Note 3*: Physician statement presence or absence of adenopathy should be used.

**Note 4**: Adenopathy = presence of LNs >1.5 cm on PE and part of staging criteria.

*Note 5*: Physical exam alone. Use CT only if exam cannot be used to detect adenopathy.

*Note 6*: No mention (present or absent), code 9.

*Note 7*: Physician's stated Rai stage takes priority when conflicting info.

Description	Code
Adenopathy not identified No LN >1.5cm Physician Rai stage 0	0
Adenopathy present LN >1.5cm Physician Rai stage 1	1
NOT applicable – primary C421	5
Unknown; Not documented; No Rai staging documented <b>AND</b> adenopathy not documented; Physician Rai stage 2-4	9

### Organomegaly

**Note 1**: All cases CLL/SLL require Lugano classification, captured in AJCC stage group and 5 components of modified RAI staging system.

*Note 2*: Rai stage only applies to CLL, C421. See code 5.

*Note 3*: Physician statement presence or absence of organomegaly should be used

*Note 4*: Organomegaly = presence of enlarged liver and/or spleen on PE

*Note 5*: Physical exam alone

*Note 6*: No mention (present or absent), code 9. Both liver and spleen must be eval/normal to code 0. If only one is eval/normal, code 9.

*Note 7*: Physician Rai stage takes priority when conflicting info.

Code	Description
0	Neither hepatomegaly (liver) or splenomegaly (spleen) present. Stated Rai Stage 0-1 by physician
1	Hepatomegaly (liver) and/or splenomegaly (spleen) present. Rai stage II
5	Not applicable: Pri Site not C421
9	Not documented in record. Organomegaly not assessed, unknown if assessed. [no mention]

### Anemia

**Note 1**: All cases CLL/SLL require Lugano classification, captured in AJCC stage group and 5 components of modified RAI staging system.

*Note 2*: Rai stage only for CLL, site C421. See code 5.

**Note 3**: Anemia = Hgb < 11.0 g/dL and part of staging criteria. Use cut points in table.

*Note 4*: Record based on CBC at dx (pre-treatment). Phys statement may be used in absence of lab report.

*Note 5*: No mention of anemia or lab results, code 9.

*Note 6*: Physician stated Rai takes priority when conflicting info.

Code	Description
0	Anemia not present; Hgb >= 11.0 g/dL; stated Rai stage 0-II by physician
1	Anemia present; Hgb < 11.0 g/dL
5	Not applicable: Pri Site not C421
6	Lab value unknown, phys states patient is anemic Rai Stage III by physician
7	Test ordered, results not in chart
9	Not documented in record. Anemia not assessed, unknown if assessed. [no mention]

### Thrombocytopenia

**Note 1**: All cases CLL /SLL require Lugano classification, captured in AJCC stage group and 5 components of modified RAI staging system.

*Note 2*: Rai Stage only for CLL and pri site C421. See **code 5**.

*Note 3*: Thrombocytopenia = platelets <100,000. Part of RAI, not AJCC.

*Note 4*: Record this data item based on blood test (CBC, diff) performed at dx and pre-rx. In absence of lab test use phys statement.

*Note 5*: No mention of thrombocytopenia or lab results, code 9.

*Note 6*: Physician stated Rai takes priority when conflicting info

Code	Description
0	Thrombocytopenia not present; Platelets (PLT) >= 100,000/ul; Physician stage Rai 0-III
1	Thrombocytopenia present Platelets (PLT) <100,000 ul
5	Not applicable: Pri Site not C421
6	Lab value unkn, phy states thrombocytopenia present; Phys Rai stage IV
7	Test ordered, results not in chart
9	Not documented in record. Thrombocytopenia not assessed, unknown if assessed. [no mention]

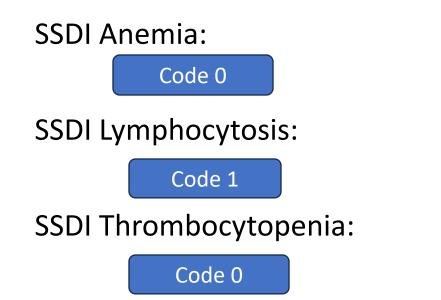
#### Case Example – CLL/SLL

Component	Value	Flag	Ref Range	Units
WBC Count	12.6	High	3.7 - 10.5	K/MM3
RBC Count	4.81		4.50 - 6.20	M/MM3
Hemoglobin	14.6		13.2 - 17.7	g/dL
Hematocrit	42		40 - 52	%
MCV (Mean Corpuscular Volume)	88		82 - 99	FL
MCH (Mean Corpuscular Hemoglobin)	30		25 - 35	PG
MCHC (Mean Corpuscular Hemoglobin Concentration)	34		32 - 36	%
Platelet Count	237		150 - 400	к/ммз



Component	Value	Flag	Ref Range	Units
% Neutrophils - Manual Diff	33.0			%
# Neutrophils - Manual Diff	4,150		2,188 - 7,800	/MM3
% Lymphocytes - Manual Diff	61.7			%
# Lymphocytes - Manual Diff	7,754	H	875 - 3,300	/MM3

#### Case Example – CLL/SLL



Component	Value	Flag	Ref Range
Hemoglobin	14.6		13.2 - 17.7
Platelet Count	237		150 - 400
# Lymphocytes - Manual Diff	7,754	Н	875 - 3,300

### Primary cutaneous lymphoma

Schema ID 00812

### EOD Primary Tumor

Code	Description	SS2018 T
100	Solitary lesion Solitary skin involvement	L
200	Regional skin involvement Multiple lesions confined to one or two contiguous body regions	RE
400	Multiple lesions involving 2 noncontiguous body regions	D
500	Multiple lesions involving ≥3 body regions	D
600	Generalized skin involvement Multiple lesions confined to three or more contiguous body regions Multiple lesions confined to discontiguous body regions	D
700	Multiple lesions, NOS	RE
800	No evidence of primary tumor	U
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in patient record Death Certificate Only	U

### EOD Regional Nodes

Code	Description	SS2018 N
000	No regional lymph node involvement	NONE
300	Peripheral node region that drains an area of current or prior skin involvement	RN
400	Two or more peripheral node regions OR involvement of any lymph node region that does not drain an area of current or prior skin involvement	RN
500	Central nodes	RN
800	Regional lymph node(s), NOS Lymph node(s), NOS	RN
999	Unknown; regional lymph node(s) not stated Regional lymph node(s) cannot be assessed Not documented in medical record Death Certificate Only	U

#### EOD Mets

Code	Description	SS2018 M
00	No distant metastases Unknown if distant metastases	NONE
10	Distant lymph node(s), NOS	D
70	Distant site(s) involved   Blood/peripheral blood  Bone marrow  Extracutaneous non-lymph node disease present  Visceral (non-cutaneous) metastasis  Distant metastasis WITH or WITHOUT distant lymph node(s)  Carcinomatosis  Distant metastasis, NOS	D
99	Death Certificate Only	U

# Plasma Cell Myeloma

Schema: 00821

# Plasma Cell Neoplasm

- Umbrella term that includes:
  - MGUS
  - Plasma cell myeloma
  - Solitary plasmacytoma
  - Extraosseous plasmacytoma
  - Osteosclerotic myeloma
- Plasma cell neoplasm only is <u>not</u> reportable
- Plasma cell neoplasm c/w multiple myeloma is <u>reportable</u>

# Plasma cell myeloma | Coding

Schema ID 00821	Always
Tumor Size Clinical	999
Tumor Size Path	999
Tumor Size Summary	999
Reg Nodes Pos/Exam	99/99
LVI	8
EOD Pri Tumor	700
EOD Reg Nodes	888
EOD Mets	88
SS18	7
Grade Clinical/Path/yc/yp	8

# Plasma Cell Myeloma

#### Code Description

0 **Multiple Myeloma** Myeloma NOS Non-secretory myeloma Plasma cell myeloma Ultra-high risk smoldering MM Smoldering plasma cell 1 myeloma Asymptomatic plasma cell myeloma Early myeloma Evolving myeloma Other terminology describing 9 myelom Unknown terms used

#### Schema Discriminator 1

- Several terms are used to characterize plasma cell myeloma at the time of diagnosis
- Select the code based on the terminology specified by the physician in the record
  - Do **NOT** attempt to determine the correct term based on the diagnostic criteria in *AJCC Table 82.1*
  - Do **NOT** change the discriminator code if a term used later indicates progression to a more aggressive disease course
  - Plasma cell leukemia and Plasma cell myeloma diagnosed simultaneously, code 0

## Plasma cell myeloma - SSDI

Schema ID 00821	NAACCR
Serum Beta-2 Microglobulin Pretreatment Level	#3931
Serum Albumin Pretreatment Level	#3930
High Risk Cytogenetics	#3857
LDH Pretreatment Level	#3869

Serum Beta-2 Microglobulin Pretreatment level **Description**: protein that is found on the surface of many cells and plentiful on the surface of white blood cells. Increased production or destruction of these cells causes Serum β2 (beta-2) Microglobulin level to increase.

Rationale: prognostic factor required in AJCC staging

- **Note 1**: part of RISS. Use cut points in table regardless of lab reference ranges.
- Note 2: Blood test performed at dx (pre-treatment). Use phys statement if no lab test. Use highest stated value.
- Note 3: No mention, code 9
- *Note 4*: If schema discriminator coded to 1 or 9, **code 5**.

# Serum Beta Microglobulin Pre-Rx Level

Codes	Description
0	β2-microblobulin < 3.5 mg/L
1	β2-microblobulin >= 3.5 g/dL <5.5 mg/L
2	β2-microblobulin >=5.5 mg/L
5	Schema Discriminator 1: Plasma Cell Myeloma term coded 1 or 9
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

## Case Example – Multiple Myeloma 9732

#### BETA 2 MICROGLOBULIN (pre-treatment results)

• Results Abnormal

Test	Value	Flag	Ref Range	Units	Status
Beta-2 Microglob	6.0	High	1.1-2.4	mg/L	Final

What would you code the SSDI – Beta-2 Microglobulin?

Codes	Description
0	β2-microblobulin < 3.5 mg/L
1	β2-microblobulin >= 3.5 g/dL <5.5 mg/L
2	β2-microblobulin >=5.5 mg/L
5	Schema Discriminator 1: Plasma Cell Myeloma term coded 1 or 9
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### Serum Albumin Pretreatment

**Description**: Albumin is the most abundant protein in human blood plasma. Serum albumin pretreatment level is a prognostic factor for plasma cell myeloma. Source: blood test.

Rationale: prognostic factor required in AJCC Staging

- Note 1: Elevated serum albumin is >=3.5 g/dL, part of RISS.
  - Use cut points in table regardless of lab's reference range.
  - Lab value stated as 3.5 g/dL is = to 35 g/L
- **Note 2**: Blood test preformed pre-treatment. Do not use urine findings. Use physician statement if no blood test.
- Note 3: No mention of serum albumin, code 9
- Note 4: If schema discriminator 1 is coded 1 or 9, code 5.

### Serum Albumin Pre-Treatment

\*A lab value expressed in grams per liter (g/L) is 10x same value expressed in g/dL

Codes	Description
0	Serum albumin < 3.5 g/dL {or *35 g/L}
1	Serum albumin >= 3.5 g/dL
5	Schema Discriminator 1: Plasma Cell Myeloma Term coded 1 or 9.
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### Case Example – Multiple Myeloma 9732

#### SERUM PROTEIN ELECTROPHORESIS

SPEP-Path Interpretation: A monoclonal protein is present in the gamma region comprising 0.3 g/dL of the total

protein.

Component	Value	Flag	Ref Range	Units	Status
SPEP- Total Protein	6.6		6.0-8.0	g/dL	Final
SPEP-Albumin	3.6	L	4.2-5.2	g/dL	Final
SPEP-Alpha1 Fraction	0.4		0.3-0.5	g/dL	Final
SPEP-Alpha2 Fraction	0.8	н	0.3-0.6	g/dL	Final
SPEP-Beta1 Fraction	0.4			g/dL	Final
SPEP-Beta2 Fraction	0.4			g/dL	Final
SPEP-Beta Fraction Total	0.8		0.6-1.0	g/dL	Final
SPEP-Gamma Fraction	0.9		0.5-1.3	g/dL	Final
SPEP-Monoclonal Protein	0.3			g/dL	Final

#### What is the correct code for Serum Albumin?

Codes	Description
0	Serum albumin < 3.5 g/dL {or *35 g/L}
1	Serum albumin >= 3.5 g/dL
5	Schema Discriminator 1: Plasma Cell Myeloma Term coded 1 or 9.
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### High Risk Cytogenetics

**Description**: one or more of <u>t(4;14), t(14;16), or del 17p</u> identified from **FISH test results** and is part of the staging criteria for plasma cell myeloma.

*Rationale*: prognostic factor required in AJCC 8th edition

- **Note 1**: Physician statement of presence or absence can be used
- **Note 2**: Based on physician statement or **FISH** interpretation (pretreatment)
- **Note 3**: If presence/absence differs from physician statement, physician's statement takes precedence.
- Note 4: If no mention, code 9
- Note 5: If schema Discriminator 1 is coded to 1 or 9, code 5.

### **High Risk Cytogenetics**



Codes	Description
0	High-risk cytogenetics not identified/not present
1	High-risk cytogenetics present "del 17p, t(4;14), or (t:14;16)"
5	Schema Discriminator 1: Plasma Cell Myeloma Term coded 1 or 9
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### Case Example 1 – Multiple Myeloma 9732

#### **FISH Final Result**

Clinical Indication: MM PANEL

Results/Interpretation NORMAL FISH RESULT

- Probe: Normal range Result (% abnormal nuclei)
- CKS1B/p18 [1q21/1p32] Gain of CKS1B <1.5%
  - Loss of p18 <5.6% normal
- TAS2R1/CEP9/CEP15 [5p15.31/9CEN/15CEN]
  - Gain of 5 < 1%
  - Gain of 9 < 1.2%
  - Gain of 15 <1%
- D13S319/13q34 [13q14.3/13q34] 13q deletion <4.4%
  - Monosomy 13: <4% normal
- IgH [14q32.3]
  - IgH rearrangement <1% normal
- p53/CEP10 [17p13.1/10cen]
  - Loss of p53 <3.7% normal

All probe sets exhibited normal signal patterns.

### What is the correct code for High Risk Cytogenetics?

Codes	Description
0	High-risk cytogenetics not identified/not present
1	High-risk cytogenetics present "del 17p, t(4;14), or (t:14;16)"
5	Schema Discriminator 1: Plasma Cell Myeloma Term coded 1 or 9
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### **Case Example 2**

Clinical Indication: MM PANEL, MULTIPLE MYELOMA

- Results/Interpretation:
   ABNORMAL FISH RESULTS
  - Polysomy of chromosome 5, 9 and 15 are associated with plasma cell disorders, including multiple myeloma.

Probe	Normal range	Result (% abnormal nuclei)
CKS1B/p18 [1q21/1p32]	Gain of CKS1B <1.5% Loss of p18 <5.6%	normal
TAS2R1/CEP9/CEP15 [5p15.31/9CEN/15CEN]	Gain of 5 <1% Gain of 9 <1.2% Gain of 15 <1%	Polysomy including trisomy 5, 9, 15 or tetrasomy 15 (87%)
D13S319/13q34 [13q14.3/13q34]	13q deletion <4.4% Monosomy 13 <4%	normal
lgH [14q32.3]	lgH rearrangement <1%	normal
p53/CEP10 [17p13.1/10cen]	Loss of p53 <3.7%	No deletion, one extra p53 signal (89%) likely from hyperdiploid clone

What is the correct code for High Risk Cytogenetics? Code 0 No mention del 17p, t(4;14) or t(14;16)

# LDH Pretreatment level

**Description**: enzyme involved in conversion of sugars to energy and present in most cells in the body. Elevated pretreatment LDH is an adverse prognostic factor for plasma cell myeloma and melanoma of the skin.

Rationale: prognostic factor required in AJCC 8th edition

- **Note 1**: Use reference ranges from your lab to determine if LDH is normal.
- **Note 2**: Blood test performed at dx (pre-treatment). Use phys statement if no lab test. Use highest stated value.
- Note 3: No mention, code 9
- Note 4: If schema discriminator 1 is coded to 1 or 9, code 5.

### **LDH Pretreatment Level**

Codes	Description
0	Normal LDH level Low, below normal
1	Above normal LDH level; High
5	Schema Discriminator 1: Plasma Cell Myeloma term coded 1 or 9
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### Case Example – Multiple Myeloma 9732

#### • LACTATE DEHYDROGENASE:

Component	Value	Ref Range	Units	Status
LDH	162	135-225	U/L	Final
		162 is within normal range		

#### What is the correct code for LDH?

Codes	Description
0	Normal LDH level Low, below normal
1	Above normal LDH level; High
5	Schema Discriminator 1: Plasma Cell Myeloma term coded 1 or 9
7	Test ordered, result not in chart
9	Not documented in medical record, not assessed or unknown if assessed

### MM Surgery Codes

Always	Code
Surgery Primary Site	A980
Scope Reg LNs	9
Surg Other/Reg/Dist	

**Surgery Codes** 

Hematopoietic/Reticuloendothelial/ Immunoproliferative/Myeloproliferative Disease C420, C421, C423, C424 (with any histology)

#### Codes

A980 All hematopoietic/reticuloendothelial/immunoproliferative/myeloproliferative disease sites and/or histologies, WITH or WITHOUT surgical treatment

Surgical procedures for hematopoietic, reticuloendothelial, immunoproliferative, myeloproliferative primaries are to be recorded using the data item *Surgical Procedure of Other Site* (NAACCR #1294).

[SEER Note: A990 Death certificate only.]

# Plasma Cell Disorders

Schema 00822

## **Plasma Cell Disorders**

Primary Site	Histology
C000-C440, C442-C689, C691-C694, C698-C699, C739-C750, C754-C809	9671,9734
C000-C699, C739-C750, C754-C809	9731, 9761
C700-C729, C751-C753	9731, 9734, 9761
C700-C729, C751-C753	9671

#### Notes

9671 Lymphoplasmacytic lymphoma (except C441, C690, C695-C696)

> C700-C729, C751-C753 (2018-2022 only) (See Note 2)

9731 Plasmacytoma, NOS

9734 Plasmacytoma, extramedullary (except C441, C690, C695-C696)

9761 Waldenstrom macroglobulinemia

# Plasmacytoma

#### Solitary plasmacytoma, medullary 9731/3

- Occurs in bone
- Single plasmacytoma in the bone (not multiple bone involvement)
- No bone marrow involvement

#### Plasmacytoma, extramedullary 9734/3

- Occurs outside of the bone
- Primary site not in bone
- No bone marrow involvement

#### Multiple plasmacytomas – occurring in bone or outside

• Code as Plasma Cell Myeloma – 9732/3

# Lymphoplasmacytic Lymphoma

#### Lymphoplasmacytic Lymphoma 9671/3

- Common sites: LN, bone marrow, and other organs
- Diagnosed by an increased number of immunoglobulins
- Module 6: PH17 and Module 7

#### Waldenstrom Macroglobulinemia (WM) 9761/3

- Primary site dependent on diagnosis year:
- 2010-2017 code C420 (blood)
- 2018+ code C421 (bone marrow)
- A subset of lymphoplasmacytic lymphoma
- Module 6: PH16

#### Gamma Heavy Chain Disease 9762/3

- Variant of LPL
- Heavy chain deposition (HCD) is NOT the same disease HCD is non-reportable
- Module 7

Description	EOD Primary Tumor	SS18
Single plasmacytoma occurring in bone – <b>9731</b> Single plasmacytoma NOS – <b>9731</b> WITH or WITHOUT soft tissue extension	100	L
Single plasmacytoma – <b>9734</b> Single plasmacytoma occurring outside of bone – <b>9734</b>	200	L
Lymphoplasmacytic lymphoma – <b>9671</b> Waldenstrom Macroglobulinemia – <b>9761</b>	700	D
Unknown Primary tumor can't be assessed Not documented DCO	999	U

### Plasma Cell Disorders – EOD Primary Tumor

### Plasma Cell Disorder – EOD Reg LN & Mets

Description	EOD Reg LN	SS18
No regional LN involvement	000	None
Extraosseous plasmacytomas <u>ONLY</u> – 9734 • Regional LN NOS; LN NOS	800	RN
<ul> <li>Not applicable:</li> <li>Lymphoplasmacytic lymphoma – 9671</li> <li>Plasmacytoma – 9731</li> <li>Single plasmacytoma occurring in bone – 9731</li> <li>Waldenstrom Macroglobulinemia – 9761</li> </ul>	987	NA
Unknown Reg LN not stated Reg LN can't be assessed Not documented DCO	999	U

EOD Mets: Code 88 Not Applicable for all histologies

### Plasma Cell Disorder – Summary Stage 18

Description	Note	SS18 Code
<ul> <li>Single plasmacytoma occurring in bone – 9731</li> <li>WITH or WITHOUT soft tissue extension</li> <li>Single plasmacytoma NOS – 9734</li> <li>Occurring outside bone</li> </ul>	Localized	1
<ul> <li>Regional LN involved only</li> <li>Extraosseous plasmacytoma only – 9734</li> <li>Regional LN NOS</li> <li>LN NOS</li> </ul>	Reg LN	3
<ul> <li>Distant site(s)/lymph nodes involved</li> <li>Lymphoplasmacytic lymphoma – 9671</li> <li>Plasma cell myeloma – 9732</li> <li>Waldenstrom Macroglobulinemia – 9761</li> <li>Multiple osseous or medullary plasmacytomas</li> <li>Multiple plasmactyomas NOS</li> </ul>	Distant	7
<ul><li>Unknown if extension or mets</li><li>Applicable for 9731 and 9734 ONLY</li></ul>	Unknown	9

## Transformation

- Multiple Primary Rules:
  - Exception for plasmacytoma (9731 & 9734) and plasma cell myeloma (9732)
    - M10
      - This rule would only apply in the initial workup was complete and a <u>single plasmacytoma</u> was diagnosed <u>AND</u>
      - Plasma cell myeloma is diagnosed <u>after workup and treatment</u>, then this <u>rule would apply</u> and be a second primary
    - M11
      - Rule does **NOT** apply plasmacytoma and plasma cell myeloma diagnosed simultaneously or during initial workup is a single primary
    - M13
      - Rule does **NOT** apply plasmacytoma after diagnosis of plasma cell myeloma is a single primary

# SEER\*Educate

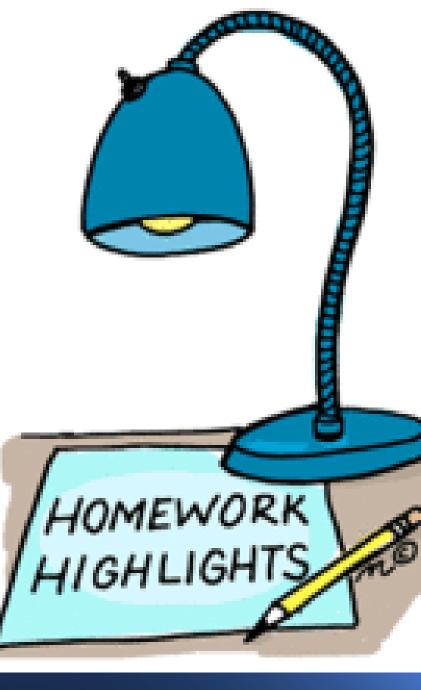
#### Training | Coding CEs Dx 2021-2024 EOD, Summary Stage, Grade, SSDI Mashup

O Lymphoma-CLL/SLL

Cases 01-05

 $\circ$  Hodgkin & Non-Hodgkin Lymphoma

• Cases 01-05



# Questions

#### **Contact Info**

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