HEMATOLOGIC MALIGNANCIES
LYMPH NODES AND LYMPHATIC STRUCTURES ABOVE AND BELOW THE DIAPHRAGM

**Note**: The most common terms are included in this table. For a more complete listing of lymph nodes, see Appendix I.

<table>
<thead>
<tr>
<th>ABOVE THE DIAPHRAGM</th>
<th>BELOW THE DIAPHRAGM</th>
</tr>
</thead>
<tbody>
<tr>
<td>Axillary</td>
<td>Celiac</td>
</tr>
<tr>
<td>Brachial (lateral axillary)</td>
<td>Colic</td>
</tr>
<tr>
<td>Buccal (buccinator)</td>
<td>Femoral (superficial, inguinal)</td>
</tr>
<tr>
<td>Cervical, NOS</td>
<td>Gastric</td>
</tr>
<tr>
<td>Epitrochlear</td>
<td>Hepatic</td>
</tr>
<tr>
<td>Facial</td>
<td>Ileocolic</td>
</tr>
<tr>
<td>Hilar (bronchopulmonary)</td>
<td>Iliac</td>
</tr>
<tr>
<td>Infraclavicular (subclavicular)</td>
<td>Inguinal</td>
</tr>
<tr>
<td>Internal jugular</td>
<td>Internal iliac (hypogastric)</td>
</tr>
<tr>
<td>Mastoid (post-/retro-auricular)</td>
<td>Mesenteric</td>
</tr>
<tr>
<td>Mediastinal</td>
<td>Obturator</td>
</tr>
<tr>
<td>Occipital</td>
<td>Pancreatic</td>
</tr>
<tr>
<td>Para/peritralcar</td>
<td>Para-aortic</td>
</tr>
<tr>
<td>Parasternal (internal mammary)</td>
<td>Peyer’s patches</td>
</tr>
<tr>
<td>Parotid</td>
<td>Popliteal</td>
</tr>
<tr>
<td>Pectoral (anterior axillary)</td>
<td>Porta hepatis (portal)</td>
</tr>
<tr>
<td>Posterior triangle (spinal accessory)</td>
<td>Retroperitoneal</td>
</tr>
<tr>
<td>Preauricular</td>
<td>Sacral</td>
</tr>
<tr>
<td>Prelaryngeal</td>
<td>Spleen</td>
</tr>
<tr>
<td>Retropharyngeal</td>
<td>Splenic (lineal)</td>
</tr>
<tr>
<td>Scalene (inferior deep cervical)</td>
<td></td>
</tr>
<tr>
<td>Sublingual</td>
<td></td>
</tr>
<tr>
<td>Submandibular (submaxillary)</td>
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<tr>
<td>Submental</td>
<td></td>
</tr>
<tr>
<td>Subscapular (posterior axillary)</td>
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</tr>
<tr>
<td>Supraclavicular (transverse cervical)</td>
<td></td>
</tr>
<tr>
<td>Tonsil</td>
<td></td>
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<tr>
<td>Thymus</td>
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</tr>
<tr>
<td>Waldeyer ring [ring of lymphoid tissue formed by the two palatine tonsils, the pharyngeal tonsil (adenoids) and the lingual tonsil]</td>
<td></td>
</tr>
</tbody>
</table>
HEMATOLOGIC MALIGNANCIES

LYMPHOMA

9590, 9596-9663, 9673-9699, 9702-9719, 9725-9726, 9735, 9737-9738, 9823, 9826-9827
(varying primary sites and histologies)

- See Summary Stage 2018 Manual, Appendix III for a detailed listing of primary site/histology combinations for this schema

9591 and Schema Discriminator 1: 3, 9 (C000-C440, C442-C689, C691-C694, C698-C809)

Note 1: The following sources were used in the development of this chapter

- Collaborative Stage Data Collection System, version 02.05: https://cancerstaging.org/cstage/Pages/default.aspx

Note 2: See the following schemas for the listed histologies

- C441, C690, C695-C696 (9590-9699, 9702-9719, 9725-9726, 9734-9738, 9823, 9826-9827, 9930): Lymphoma Ocular Adnexa
- C440, C442-C449, C510, C609, C632 (9597, 9680, 9708-9709, 9712, 9718-9719, 9726): Primary Cutaneous Lymphomas
- C700, C710-C719 (9680, 9699, 9700-9715): Brain
- C701, C709, C720-C725, C728-C729 (9680, 9699, 9700-9715): CNS Other
- C751-C753 (9680, 9699, 9700-9715): Intracranial Gland
- 9591 and Schema Discriminator 1: 1, 2 (C000-C440, C442-C689, C691-C694, C698-C809): HemeRetic

Note 3: Chapter includes the preferred terms based on the 2017 WHO Classification of Haematopoietic and Lymphoid Tissues

9590 Malignant lymphoma, NOS
9591 Non-Hodgkin lymphoma, NOS
9596 B-cell lymphoma, unclassifiable, with features intermediate between DLBCL and classic Hodgkin lymphoma
9597 Primary cutaneous follicle centre lymphoma
9650 Classical Hodgkin lymphoma
9651 Lymphocyte-rich classic Hodgkin lymphoma
9652 Mixed cellularity classic Hodgkin lymphoma
9653 Lymphocyte-depleted classic Hodgkin lymphoma
9659 Nodular lymphocyte predominant Hodgkin lymphoma
9663 Nodular sclerosis classic Hodgkin lymphoma
9673 Mantle cell lymphoma
9678 Primary effusion lymphoma
9679 Primary mediastinal (thymic) large B-cell lymphoma
9680 Diffuse large B-cell lymphoma (DLBCL)
9687 Burkitt lymphoma
9688 T-cell/histiocyte-rich large B-cell lymphoma
9689 Splenic marginal zone lymphoma
9690 Follicular lymphoma
9691 Follicular lymphoma, grade 2
9695 Follicular lymphoma, grade 1
9698 Follicular lymphoma, grade 3
9699 Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)
9702 Peripheral T-cell lymphoma, NOS
9705 Angioimmunoblastic T-cell lymphoma
9708 Subcutaneous panniculitis-like T-cell lymphoma
9709 Primary cutaneous peripheral T-cell lymphomas
9712 Intravascular large B-cell lymphoma
9714 Anaplastic large cell lymphoma, ALK-positive
9715 Anaplastic large cell lymphoma, ALK-negative (2021+ only)
9716 Hepatosplenic T-cell lymphoma
9717 Enteropathy-associated T-cell lymphoma
9718 Primary cutaneous anaplastic large cell lymphoma
9719 Extranodal NK/T-cell lymphoma, nasal type
9725 Hydroa vacciniforme-like lymphoma (2018-2020 only, nonreportable as of 2021)
9726 Primary cutaneous gamma-delta T-cell lymphoma (2018-2020 only, see code 9687/3 for 2021+)
9735 Plasmablastic lymphoma
9737 ALK-positive large B-cell lymphoma
9738 HHV8-positive DLBCL, NOS
9766 Lymphomatoid granulomatosis grade 3 (2021+ only)
9823 Chronic lymphocytic leukemia (CLL)/small lymphocytic lymphoma
9826 Burkitt cell leukemia (2018-2020 only, see code 9687/3 for 2021+)
9827 Adult T-cell leukemia/lymphoma

**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.

**Note 5:** If there is peripheral blood or bone marrow involvement, code 7.
SUMMARY STAGE

1 Localized only

Nodal Lymphomas

- Single lymph node region involved
- Involvement of multiple nodal chains in the SAME lymph node region

Extranodal Lymphomas

- Single extralymphatic site
  - WITHOUT nodal involvement (see code 2 for WITH nodal involvement)
- Multifocal involvement of one extralymphatic organ/site (EXCEPT multifocal lung involvement or any liver involvement, see code 7)
  - WITHOUT nodal involvement (see code 7 for WITH nodal involvement)

2 Regional by direct extension only

Bulky disease present

Nodal Lymphomas

- Two or more lymph node regions involved SAME side of diaphragm
- Contiguous extension between extralymphatic sites and regional nodes
  - WITH or WITHOUT involvement of other nodal regions on SAME side of diaphragm

Extranodal Lymphomas

- Localized involvement of a single extralymphatic organ/site
  - WITH involvement of its regional lymph node(s) OR
  - WITH involvement of other lymph node(s) on the SAME side of the diaphragm

7 Distant site(s)/lymph node(s) involved

- Distant involvement
  - Diffuse or disseminated involvement of ONE extralymphatic organ/site WITH or WITHOUT nodal involvement
  - Diffuse or disseminated involvement of MORE than one extralymphatic organ/site WITH or WITHOUT nodal involvement
  - Involvement of isolated extralymphatic organ in absence of involvement of adjacent lymph nodes, but in conjunction with disease in distant sites
  - Involvement of lymph node regions on BOTH sides of the diaphragm
- OR nodes ABOVE the diaphragm involved WITH spleen involvement
  - Multifocal involvement of one extralymphatic organ/site WITH nodal involvement
  - Noncontiguous extralymphatic organ involvement in conjunction with nodal disease (two or more sites involved)
- Distant metastasis, NOS
  - Blood/peripheral blood
  - Bone marrow
  - Cerebrospinal fluid (CSF)
  - Liver
  - Lung (other than by direct extension in code 2)

9 Unknown if extension or metastasis
MYCOSIS FUNGOIDES

9700-9701 (C000-C002, C006, C440-C449, C510-C512, C518-C519, C600-C602, C608-C609, C632)

Note 1: The following sources were used in the development of this chapter

- Collaborative Stage Data Collection System, version 02.05: https://cancerstaging.org/cstage/Pages/default.aspx

Note 2: For mycosis fungoides of sites other than those above, use the appropriate site-specific schema.

Note 3: Code 0 is not applicable for this chapter.

Note 4: All lymph node (regional and distant) involvement is coded as regional lymph node involvement.

Note 5: Visceral involvement is metastatic disease and should be questioned in the absence of node or blood involvement.

SUMMARY STAGE

1 Localized only (localized, NOS)

- MFCG Stage I
  - Less than 10% of skin surface, no tumors
  - Limited to patches, papules, or plaques
- MFCG Stage II
  - Greater than or equal to 10% of skin surface, no tumors
  - Generalized patches, papules, or plaques
- Not stated whether patches, papules, or plaques
  - % of body surface not stated, no tumors
  - Skin involvement, NOS: extent not stated, no tumors
2 Regional by direct extension only

- MFCG Stage III
  - Cutaneous tumor, size not stated
  - Generalized erythroderma (confluence of erythema)
    - (greater than 50% of body involved with diffuse redness)
  - One or more tumors equal to 1 cm or greater
  - Sezary syndrome
  - Skin lesion described as tumor less than 1 cm

3 Regional lymph node(s) involved only

- Both clinically enlarged palpable lymph node(s) (adenopathy) AND
  - pathologically positive nodes
- Clinically enlarged palpable lymph node(s) (adenopathy) AND
  - either pathologically negative nodes or no pathological statement
- No clinically enlarged palpable lymph node(s) (adenopathy) BUT
  - pathologically positive lymph node(s)
- Dutch grade 1-4 OR NCI LN 0-4
  - Clone positive, negative or unknown
- Regional lymph node(s), NOS
  - Lymph node(s), NOS

4 Regional by BOTH direct extension AND regional lymph node(s) involved

- Codes (2) + (3)

7 Distant site(s) involved

- Distant site(s) (including further contiguous extension)
  - MFCG Stage IV
    - Bone marrow
    - Involvement by at least one organ outside the skin, nodes, blood, or bone marrow
    - Liver
    - Spleen
    - Visceral (non-cutaneous, extranodal) involvement, pathologically confirmed
- Distant metastasis, NOS
  - Carcinomatosis

9 Unknown if extension or metastasis
PRIMARY CUTANEOUS LYMPHOMAS (EXCLUDING MF AND SS)

9597, 9680, 9708-9709, 9712, 9718-9719, 9726
C440, C442-C449, C510, C609, C632
C440 Skin of lip, NOS
C442 External ear
C443 Skin of other and unspecified parts of face
C444 Skin of scalp and neck
C445 Skin of trunk
C446 Skin of upper limb and shoulder
C447 Skin of lower limb and hip
C448 Overlapping lesion of skin
C449 Skin, NOS
C510 Labium majus
C609 Penis
C632 Scrotum, NOS

Note 1: The following sources were used in the development of this chapter

- Collaborative Stage Data Collection System, version 02.05: https://cancerstaging.org/cstage/Pages/default.aspx

Note 2: Code 0 is not applicable for this chapter.

Note 3: See the Mycosis Fungoides chapter for Mycosis Fungoides (9700) and Sezary syndrome (9701).

Note 4: Chapter includes the preferred terms based on the 2017 WHO Classification of Haematopoietic and Lymphoid Tissues

9597 Primary cutaneous follicle center lymphoma
9680 Primary cutaneous diffuse large B-cell lymphoma, leg type
9708 Subcutaneous panniculitis-like T-cell lymphoma
9709 Primary cutaneous peripheral T-cell lymphomas
9712 Intravascular large B-cell lymphoma
9718 Primary cutaneous anaplastic large cell lymphoma
9719 Extranodal NK/T-cell lymphoma, nasal type
9726 Primary cutaneous gamma-delta t-cell lymphoma
Note 5: If there is peripheral blood or bone marrow involvement, code 7.

SUMMARY STAGE

1 Localized only
   - Solitary lesion
   - Solitary skin involvement

2 Regional by direct extension only
   - Multiple lesions confined to one or two contiguous body regions
   - Multiple lesions, NOS
   - Regional skin involvement

3 Regional lymph node(s) involved only
   - Central
   - Peripheral node region that drains an area of current or prior skin involvement
   - Regional lymph node(s), NOS
     - Lymph node(s), NOS

4 Regional by BOTH direct extension AND regional lymph node(s) involved
   - Codes (2) + (3)

7 Distant site(s)/lymph node(s) involved
   - Distant site(s) (including further contiguous extension)
     - Blood/peripheral blood
     - Bone marrow
     - Extracutaneous non-lymph node disease present
     - Generalized skin involvement
     - Multiple lesions confined to discontiguous body regions
     - Multiple lesions confined to three or more contiguous body regions
     - Visceral (non-cutaneous) metastasis
   - Distant lymph node(s), NOS
   - Distant metastasis, NOS
     - Carcinomatosis
     - Distant metastasis WITH or WITHOUT distant lymph node(s)

9 Unknown if extension or metastasis
MYELOMA AND PLASMA CELL DISORDERS

9671 Lymphoplasmacytic lymphoma (except C441, C690, C695-C696)
9731 Plasmacytoma, NOS
9732 Plasma cell myeloma/multiple myeloma
9734 Plasmacytoma, extramedullary (except C441, C690, C695-C696)
9761 Waldenstrom Macroglobulinemia

Note 1: The following sources were used in the development of this chapter

- Collaborative Stage Data Collection System, version 02.05: https://cancerstaging.org/cstage/Pages/default.aspx

Note 2: Codes 0, 2, and 4 are not applicable for this chapter.

Note 3: Plasma cell myeloma/multiple myeloma (9732) is a widely disseminated plasma cell neoplasm, characterized by a single clone of plasma cells derived from B cells that grows in the bone marrow. It is always coded to 7 for systemic involvement.

Note 4: Lymphoplasmacytic lymphoma (9671) and Waldenstrom Macroglobulinemia (9761) are now collected with the plasma cell disorders. These are systemic diseases and should always be coded 7.

SUMMARY STAGE

1 Localized only

- Single plasmacytoma occurring in bone (osseous or medullary) (9731)
  - WITH or WITHOUT soft tissue extension
- Single plasmacytoma, NOS (9734)
  - Single plasmacytoma occurring outside of bone (extraosseous or extramedullary) (9731)

3 Regional lymph node(s) involved only

- Extraosseous plasmacytomas only (9734)
  - Regional lymph node(s), NOS
    - Lymph node(s), NOS
7 Distant site(s)/lymph node(s) involved

- Lymphoplasmacytic lymphoma (9671)
- Plasma cell myeloma (9732)
  - Multiple myeloma
  - Myeloma, NOS
  - Multiple extraosseous or extramedullary plasmacytomas
  - Multiple osseous or medullary plasmacytomas
  - Multiple plasmacytomas, NOS
- Waldenstrom Macroglobulinemia (9761)

9 Unknown if extension or metastasis (applicable for 9731 and 9734 only)
HEMERETIC

9724, 9727, 9740-9742, 9749, 9762-9809, 9811-9820, 9831-9920, 9931-9993

9591 and Schema Discriminator 1: 1, 2 (C000-C440, C442-C689, C691-C694, C698-C809)

9751, 9755-9759 (C000-C699, C739-C750, C754-C809)

9930 (C000-C440, C442-C689, C691-C694, C698-C809)

Note 1: The following sources were used in the development of this chapter

- Collaborative Stage Data Collection System, version 02.05: https://cancerstaging.org/cstage/Pages/default.aspx

Note 2: See the following schemas for the listed histologies

- 9591 and Schema Discriminator 1: 3, 9 (C000-C440, C442-C689, C691-C694, C698-C809): Lymphoma
- 9751, 9755-9759 (C700, C710-C719): Brain
- 9751, 9755-9759 (C701, C709, C720-C725, C728-C729): CNS Other
- 9751, 9755-9759 (C751-C753): Intracranial Gland
- 9930 (C441, C690, C695-C696): Lymphoma Ocular Adnexa

Note 3: The following histologies can be localized (code 1), systemic (7) or unknown (9)

9740 Mast cell sarcoma
9749 Erdheim-Chester disease (2021+ only)
9751 Langerhans cell histiocytosis, disseminated (except C700-C729, C751-C753)
9755 Histiocytic sarcoma (except C700-C729, C751-C753)
9756 Langerhans cell sarcoma (except C700-C729, C751-C753)
9757 Interdigitating dendritic cell sarcoma (except C700-C729, C751-C753)
9758 Follicular dendritic cell sarcoma (except C700-C729, C751-C753)
9759 Fibroblastic reticular cell tumor (except C700-C729, C751-C753)
9930 Myeloid sarcoma (except C441, C690, C695-C696)
9971 Polymorphic PTLD (2018-2020 only, nonreportable as of 2021)

Note 4: For histologies listed in Note 2, it is possible to have lymph node involvement; however, at this time, lymph node involvement for these histologies is not collected.
Note 5: The following histologies are systemic (code 7):

9591 Splenic B-cell lymphoma/leukemia, unclassifiable (except C441, C690, C695-C696)
9724 Systemic EBV-positive T-cell lymphoma of childhood
9727 Blastic plasmacytoid dendritic cell neoplasm
9741 Systemic mastocytosis with an associated hematological neoplasm
9742 Mast cell leukemia
9762 Heavy chain diseases
9800 Leukemia, NOS
9801 Acute undifferentiated leukemia
9806 Mixed-phenotype acute leukemia with t(9;22)(q34.1;q11.2); BCR-ABL1
9807 Mixed-phenotype acute leukemia with t(v;11q23.3); KMT2A-rearranged
9808 Mixed-phenotype acute leukemia, B/myeloid, NOS
9809 Mixed-phenotype acute leukemia, T/myeloid, NOS
9811 B-lymphoblastic leukemia/lymphoma, NOS
9812 B-lymphoblastic leukemia/lymphoma with t(9;22)(q34.1;q11.2); BCR-ABL1
9813 B-lymphoblastic leukemia/lymphoma with t(v;11q23.3); KMT2A-rearranged
9814 B-lymphoblastic leukemia/lymphoma with t(12;21)(p13.2;q22.1); ETV6-RUNXI
9815 B-lymphoblastic/lymphoma with hyperdiploidy
9816 B-lymphoblastic/lymphoma with hypodiploidy (hypodiploid ALL)
9817 B-lymphoblastic/lymphoma with t(5;14)(q31.1;q32.1); IGH/IL3
9818 B-lymphoblastic/lymphoma with t(1;19)(q23;p13.3); TCF3-PBX1
9819 B-lymphoblastic/lymphoma, BCR-ABL1-like (2021+ only)
9820 Lymphoid leukemia, NOS
9831 T-cell large granular lymphocytic leukemia
9832 Prolymphocytic leukemia, NOS
9833 B-cell prolymphocytic leukemia
9834 T-cell prolymphocytic leukemia
9837 T-lymphoblastic leukemia/lymphoma
9840 Pure erythroid leukemia
9860 Myeloid leukemia, NOS
9861 Acute myeloid leukemia, NOS
9863 Chronic myeloid leukemia
9865 Acute myeloid leukemia with t(6;9)(p23;q34.1); DEK-NUP214
9866 Acute promyelocytic leukemia with PML-RARA
9867 Acute myelomonocytic leukemia
9869 Acute myeloid leukemia with inv(3)(q21.3q26.2) or t(3;3)(q21.3;q26;2); RBM15-MKL1
9870 Acute basophilic leukemia
9871 Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFB-MYH11
9872 Acute myeloid leukemia, minimal differentiation
9873 Acute myeloid leukemia without maturation
9874 Acute myeloid leukemia with maturation
9875 Chronic myeloid leukemia, BCR-ABL1-positive
9876 Atypical chronic myeloid leukemia BCR-ABL1-negative
9877 Acute myeloid leukemia with mutated NPM1 (2021+ only)
9878 Acute myeloid leukemia with biallelic mutation of CEBPA (2021+ only)
9879 Acute myeloid leukemia with mutated RUNX1 (2021+ only)
9891 Acute monoblastic and monocytic leukemia
9895 Acute myeloid leukemia with myelodysplasia-related changes
9896 Acute myeloid leukemia with t(8;21)(q22;q22.1), RUNX1-RUNX1T1
9897 Acute myeloid leukemia with t(9;11)(p21.3;q23.3); KMT2A-MLLT3
9898 Myeloid leukemia associated with Down Syndrome
9910 Acute megakaryoblastic leukemia
9911 Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13.3;q13.1); RBM15-MKL1
9912 Acute myeloid leukemia with BCR-ABL1 (2021+ only)
9920 Therapy-related myeloid neoplasms
9931 Acute panmyelosis with myelofibrosis
9940 Hairy cell leukemia
9945 Chronic myelomonocytic leukemia, NOS
9946 Juvenile myelomonocytic leukemia
9948 Aggressive NK-cell leukemia
9950 Polycythemia vera
9961 Primary myelofibrosis
9962 Essential thrombocythemia
9963 Chronic neutrophilic leukemia
9964 Chronic eosinophilic leukemia, NOS
9965 Myeloid/lymphoid neoplasms with PDGFRA rearrangement
9966 Myeloid/lymphoid neoplasm with PDGFRB rearrangement
9967 Myeloid/lymphoid neoplasm with FGFR1 rearrangement
9968 Myeloid/lymphoid neoplasm with PCM1-JAK2 (2021+ only)
9975 Myelodysplastic/myeloproliferative neoplasm, unclassifiable
9980 Myelodysplastic syndrome with single lineage dysplasia
9982 Myelodysplastic syndrome with ring sideroblasts and single lineage dysplasia
9983 Myelodysplastic syndrome with excess blasts
9985 Myelodysplastic syndrome with multilineage dysplasia
9986 Myelodysplastic syndrome with isolated del(5q)
9989 Myelodysplastic syndrome, unclassifiable
9991 Refractory neutropenia (2018-2020 only, see code 9980/3 for 2021+)
9992 Refractory thrombocytopenia (2018-2020 only, see code 9980/3 for 2021+)
9993 Myelodysplastic syndrome with ring sideroblasts and multilineage dysplasia (2021+ only)

**Note 6:** Summary Stage is the only applicable staging system for this site/histology/schema.

**Note 7:** Codes 0, 2, 3, 4, and 5 are not applicable for this chapter.

**SUMMARY STAGE**

1 **Localized only**

- Localized disease
- (Single/solitary/unifocal/isolated)
- See Notes 2 and 3
7 Distant site(s)/lymph node(s) involved

- Systemic disease
- See Note 4

9 Unknown if extension or metastasis