

KAPOSI SARCOMA OF ALL SITES

(M-9140)

SUMMARY STAGE

1 Localized only

Single lesion or multiple lesions in ONE of the following:

Mucosa (e.g., oral cavity, anus, rectum, vagina, vulva)

Skin

Viscera (e.g., pulmonary, gastrointestinal tract, spleen, other)

2 Regional by direct extension only

Multiple lesions in any TWO of the following:

Mucosa (e.g., oral cavity, anus, rectum, vagina, vulva)

Skin

Viscera (e.g., pulmonary, gastrointestinal tract, spleen, other)

3 Lymph node(s) involved only

Both clinically enlarged palpable lymph node(s) (adenopathy) and pathologically positive lymph nodes

Clinically enlarged palpable lymph node(s) (adenopathy), and either pathologically negative nodes or no pathological statement

No clinically enlarged palpable lymph nodes(s) (adenopathy) but pathologically positive lymph node(s)

Lymph node(s), NOS

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

Codes (2) + (3)

7 Distant site(s) involved

Lesions in ALL THREE of the following:

Mucosa (e.g., oral cavity, anus, rectum, vagina, vulva)

Skin

Viscera (e.g., pulmonary, gastrointestinal tract, spleen, other)

Further contiguous extension

Metastasis

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9 Unknown if extension or metastasis

Multiple lesions, NOS

Note 1: Codes 0 and 5 are not applicable for this scheme.

Note 2: Since there was no separate staging scheme in either the Historic Stage or the 1977 Summary Staging Guide for Kaposi sarcoma, these cases would have been staged previously using the scheme for “skin other than melanoma” for cases which arose in a skin site. For cases which arose in mucosal or visceral sites, the scheme for that site would have been used to assign stage.

**LYMPH NODES AND LYMPHATIC STRUCTURES
ABOVE AND BELOW THE DIAPHRAGM**

ABOVE the Diaphragm

Axillary
Brachial (lateral axillary)
Buccal (buccinator)
Cervical, NOS
Epitrochlear
Facial
Hilar (bronchopulmonary)
Infraclavicular (subclavicular)
Internal jugular
Mastoid (post-/retro-auricular)
Mediastinal
Occipital
Para/peritracheal
Parasternal (internal mammary)
Parotid
Pectoral (anterior axillary)
Posterior triangle (spinal accessory)
Preauricular
Prelaryngeal
Retropharyngeal
Scalene (inferior deep cervical)
Sublingual
Submandibular (submaxillary)
Submental
Subscapular (posterior axillary)
Supraclavicular (transverse cervical)
Tonsil
Thymus
Waldeyer ring [ring of lymphoid tissue formed by the
two palatine tonsils, the pharyngeal tonsil (adenoids)
and the lingual tonsil]

BELOW the Diaphragm

Celiac
Colic
Femoral (superficial inguinal)
Gastric
Hepatic
Ileocolic
Iliac
Inguinal
Internal iliac (hypogastric)
Mesenteric
Obturator
Pancreatic
Para-aortic
Peyer's patches
Popliteal
Porta hepatis (portal)
Pyloric
Retroperitoneal
Sacral
Spleen
Splenic (lienal)

HODGKIN AND NON-HODGKIN LYMPHOMAS OF ALL SITES

[excluding Mycosis Fungoides and Sezary Disease of sites listed on page 176]
(M-9590-9591, 9596, 9650-9655, 9659, 9661-9665, 9667, 9670-9671, 9673, 9675, 9678-9680, 9684, 9687, 9689-9691, 9695, 9698-9702, 9705, 9708-9709, 9714, 9716-9719, 9727-9729, 9823)

SUMMARY STAGE

1 Localized

Stage I

Involvement of a single lymph node region

Stage IE

Localized involvement of a single extralymphatic organ/site

Multifocal involvement of one extralymphatic organ/site

Stage IS

Localized involvement of spleen only

5 Regional, NOS

Stage II

Involvement of two or more lymph node regions on the SAME side of the diaphragm

Stage IIE

Direct extension to adjacent organs or tissues

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

Stage IIS

Involvement of spleen PLUS lymph node(s) BELOW the diaphragm

Stage IIES

Involvement of spleen PLUS localized involvement of a single extralymphatic organ/site BELOW the diaphragm WITH/WITHOUT involvement of lymph node(s) BELOW the diaphragm

HODGKIN AND NON-HODGKIN LYMPHOMAS OF ALL SITES
[excluding Mycosis Fungoides and Sezary Disease of sites listed on page 176]
(M-9590-9591, 9596, 9650-9655, 9659, 9661-9665, 9667, 9670-9671, 9673, 9675,
9678-9680, 9684, 9687, 9689-9691, 9695, 9698-9702, 9705, 9708-9709, 9714,
9716-9719, 9727-9729)

7 Distant

Stage III

Involvement of lymph node regions on BOTH sides of the diaphragm

Stage IIIE

Involvement of an extralymphatic organ or site PLUS involvement of lymph node(s) on the OPPOSITE side of the diaphragm

Stage IIIS

Involvement of the spleen PLUS involvement of lymph node(s) ABOVE the diaphragm

Stage IIIES

Involvement of the spleen PLUS involvement of lymph node region(s) ABOVE the diaphragm PLUS involvement of a single extralymphatic organ/site on either side of the diaphragm

Involvement of the spleen PLUS a single extralymphatic organ/site

ABOVE the diaphragm WITH OR WITHOUT involvement of lymph node(s)

Stage IV

Disseminated involvement of ONE OR MORE extralymphatic organ(s)/site(s)

(Multifocal) involvement of MORE THAN ONE extralymphatic organ/site

Metastases

Bone marrow

Liver

9 Unstaged; not stated

Note 1: E = Extralymphatic means tissues excluding lymph nodes or other lymphatic structures

Note 2: S = Spleen involvement

Note 3: Lymphatic structures include thymus gland, Waldeyer ring (tonsils), Peyer's patches (small intestine) and lymphoid nodules in the appendix. Any lymphatic structure is to be considered the same as a lymph node region. Spleen is also considered a lymphatic structure but is dealt with separately in assigning stage (see note 2).

Note 4: If there is no mention of extranodal involvement but several diagnostic procedures were done, including laparotomy, interpret as no involvement.

Note 5: Involvement of adjacent soft tissue does not alter the classification.

Note 6: Codes 0, 2, 3, and 4 are not applicable for this scheme.

Note 7: For cases diagnosed 1/1/2012 and later, SS2000 Hodgkin and Non-Hodgkin Lymphomas of All Sites staging scheme is applicable to histology code 9823.

HEMATOPOIETIC, RETICULOENDOTHELIAL, IMMUNOPROLIFERATIVE, AND MYELOPROLIFERATIVE NEOPLASMS

(M-9731-9734, 9740-9742, 9750-9758, 9760-9762, 9764-9769, 9800-9801, 9805, 9820, 9823, 9826-9827, 9831-9837, 9840, 9860-9861, 9863, 9866-9867, 9870-9876, 9891, 9895-9897, 9910, 9920, 9930-9931, 9940, 9945-9946, 9948, 9950, 9960-9964, 9970, 9975, 9980, 9982-9987, 9989)

This scheme includes the following*:

9731 = Plasmacytoma, NOS	9860 = Myeloid leukemia, NOS
9732 = Multiple myeloma	9861 = Acute myeloid leukemia, NOS
9733 = Plasma cell leukemia	9863 = Chronic myeloid leukemia, NOS
9734 = Plasmacytoma, extramedullary	9866 = Acute promyelocytic leukemia
9740 = Mast cell sarcoma	9867 = Acute myelomonocytic leukemia
9741 = Malignant mastocytosis	9870 = Acute basophilic leukemia
9742 = Mast cell leukemia	9871 = Acute myeloid leukemia with abnormal marrow eosinophils
9750 = Malignant histiocytosis	9872 = Acute myeloid leukemia, minimal differentiation
9751 = Langerhans cell histiocytosis, NOS☞	9873 = Acute myeloid leukemia without maturation
9752 = Langerhans cell histiocytosis, unifocal☞	9874 = Acute myeloid leukemia with maturation
9753 = Langerhans cell histiocytosis, multifocal☞	9875 = Chronic myelogenous leukemia, BCR/ABL positive
9754 = Langerhans cell histiocytosis disseminated	9876 = Atypical chronic myeloid leukemia BCR/ABL negative
9755 = Histiocytic sarcoma	9891 = Acute monocytic leukemia
9756 = Langerhans cell sarcoma	9895 = Acute myeloid leukemia with multilineage dysplasia
9757 = Interdigitating dendritic cell sarcoma	9896 = Acute myeloid leukemia, t(8;21)(q22;q22)
9758 = Follicular dendritic cell sarcoma	9897 = Acute myeloid leukemia, 11q23 abnormalities
9760 = Immunoproliferative disease, NOS	9910 = Acute megakaryoblastic leukemia
9761 = Waldenstrom macroglobulinemia	9920 = Therapy-related acute myeloid leukemia, NOS
9762 = Heavy chain disease, NOS	9930 = Myeloid sarcoma
9764 = Immunoproliferative small intestinal disease	9931 = Acute panmyelosis with myelofibrosis
9765 = Monoclonal gammopathy of undetermined significance☞	9940 = Hairy cell leukemia
9766 = Angiocentric immunoproliferative lesion☞	9945 = Chronic myelomonocytic leukemia, NOS
9767 = Angioimmunoblastic lymphadenopathy☞	9946 = Juvenile myelomonocytic leukemia
9768 = T-gamma lymphoproliferative disease☞	9948 = Aggressive NK-cell leukemia
9769 = Immunoglobulin deposition disease☞	9950 = Polycythemia vera
9800 = Leukemia, NOS	9960 = Chronic myeloproliferative disease, NOS
9801 = Acute leukemia, NOS	9961 = Myelosclerosis with myeloid metaplasia
9805 = Acute biphenotypic leukemia	9962 = Essential thrombocythemia
9820 = Lymphoid leukemia, NOS	9963 = Chronic neutrophilic leukemia
9823 = B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma	9964 = Hypereosinophilic syndrome
9826 = Burkitt cell leukemia	9970 = Lymphoproliferative disorder, NOS^
9827 = Adult T-cell leukemia/lymphoma (HTLV-1 positive)	9975 = Myeloproliferative disease, NOS^
9831 = T-cell large granular lymphocytic leukemia☞	9980 = Refractory anemia, NOS
9832 = Prolymphocytic leukemia, NOS	9982 = Refractory anemia with sideroblasts
9833 = Prolymphocytic leukemia, B-cell type	9983 = Refractory anemia with excess blasts
9834 = Prolymphocytic leukemia, T-cell type	9984 = Refractory anemia with excess blasts in transformation
9835 = Precursor cell lymphoblastic leukemia, NOS	9985 = Refractory cytopenia with multilineage dysplasia
9836 = Precursor B-cell lymphoblastic leukemia	9986 = Myelodysplastic syndrome with 5q deletion (5q-) syndrome
9837 = Precursor T-cell lymphoblastic leukemia	9987 = Therapy-related myelodysplastic syndrome, NOS
9840 = Acute myeloid leukemia, M6 type	9989 = Myelodysplastic syndrome, NOS

☞ Usually considered of uncertain/borderline behavior

* Only preferred terms from ICD-O-3 are given

SUMMARY STAGE

1 Localized (isolated/mono-ostotic/single/solitary/unifocal for M-9731/3, 9734/3, 9750/3, 9751/3, 9752/3)

7 Distant (polyostotic); disease disseminated at diagnosis

9 Death certificate only case

Unknown

280

Note 1: Codes 0, 2, 3, 4 and 5 are not applicable for this scheme.

Note 2: Histology codes M-9731/3 and M-9734/3 may be coded as 1, 7, or 9. Histology codes M-9740/3, M-9750/3, M-9755/3, M-9756/3, M-9757/3, M-9758/3, M-9764/3, and M-9930/3 may be coded as 1, 7, 9. Histology codes M-9751/3 and M-9752/3 can only be coded 1 unless Death certificate only (code 9). All other histologies listed above can only be coded 7 unless Death certificate only (code 9).

OTHER AND ILL-DEFINED SITES, UNKNOWN PRIMARY SITE

C76.0-C76.5, C76.7-C76.8, C80.9

C42._ and C77._, Other than hematopoietic, reticuloendothelial, immunoproliferative and myeloproliferative neoplasms (page 280), Hodgkin and non-Hodgkin lymphoma (page 278), and Kaposi sarcoma (page 274).

C42.0 Blood

C42.1 Bone marrow

C42.2 Spleen

C42.3 Reticuloendothelial system, NOS

C42.4 Hematopoietic system, NOS

Other and ill-defined sites of:

C76.0 Head, face or neck, NOS

C76.1 Thorax, NOS

C76.2 Abdomen, NOS

C76.3 Pelvis, NOS

C76.4 Upper limb, NOS

C76.5 Lower limb, NOS

C76.7 Other ill-defined sites

C76.8 Overlapping lesion of ill-defined sites

Lymph nodes of:

C77.0 Head, face and neck

C77.1 Intrathoracic

C77.2 Intra-abdominal

C77.3 Axilla or arm

C77.4 Inguinal region or leg

C77.5 Pelvic

C77.8 Lymph nodes of multiple regions

C77.9 Lymph node, NOS

C80.9 Unknown primary site

9 Unknown if extension or metastasis; unstageable

Note: Codes 0, 1, 2, 3, 4, 5, 7 are not applicable for this scheme.