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
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 (lienol)
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 IIIIE
   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

This scheme includes the following*:

9731 = Plasmacytoma, NOS
9732 = Multiple myeloma
9733 = Plasma cell leukemia
9734 = Plasmacytoma, extramedullary
9740 = Mast cell sarcoma
9741 = Malignant mastocytosis
9742 = Mast cell leukemia
9750 = Malignant histiocytosis
9751 = Langerhans cell histiocytosis, NOS
9752 = Langerhans cell histiocytosis, unifocal
9753 = Langerhans cell histiocytosis, multifocal
9754 = Langerhans cell histiocytosis disseminated
9755 = Histiocytic sarcoma
9756 = Langerhans cell sarcoma
9757 = Interdigitating dendritic cell sarcoma
9758 = Follicular dendritic cell sarcoma
9760 = Immunoproliferative disease, NOS
9761 = Waldenstrom macroglobulinemia
9763 = Chronic myelogenous leukemia
9764 = Immunoproliferative small intestinal disease
9765 = Monoclonal gammopathy of undetermined significance
9766 = Angiocentric immunoproliferative lesion
9767 = Angiitoomunoblastic lymphadenopathy
9768 = T-gamma lymphoproliferative disease
9769 = Immunoglobulin deposition disease
9800 = Leukemia, NOS
9801 = Acute leukemia, NOS
9805 = Acute biphenotypic leukemia
9820 = Lymphoid leukemia, NOS
9823 = B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma
9826 = Burkitt cell leukemia
9827 = Adult T-cell leukemia/lymphoma (HTLV-1 positive)
9831 = T-cell large granular lymphocytic leukemia
9832 = Prolymphocytic leukemia, NOS
9833 = Prolymphocytic leukemia, B-cell type
9834 = Prolymphocytic leukemia, T-cell type
9835 = Precursor cell lymphoblastic leukemia, NOS
9836 = Precursor B-cell lymphoblastic leukemia
9837 = Precursor T-cell lymphoblastic leukemia
9840 = Acute myeloid leukemia, M6 type
9841 = Acute myeloid leukemia, NOS
9845 = Chronic myelomonocytic leukemia
9846 = Juvenile myelomonocytic leukemia
9848 = Aggressive NK-cell leukemia
9850 = Polycythemia vera
9851 = Reactive lymphoid hyperplasia
9852 = Reactive lymphoid hyperplasia
9856 = Myelodysplastic syndrome with 5q deletion (5q–) syndrome
9857 = Therapy-related myelodysplastic syndrome, NOS
9860 = Myeloid leukemia, NOS
9861 = Acute myeloid leukemia, NOS
9863 = Chronic myeloid leukemia, NOS
9864 = Hyperesinophilic syndrome
9865 = Myelodysplastic syndrome
9866 = Acute promyelocytic leukemia
9867 = Acute myelomonocytic leukemia
9868 = Acute myeloid leukemia, minimal differentiation
9869 = Acute myeloid leukemia without maturation
9870 = Acute basophilic leukemia
9871 = Acute myeloid leukemia with abnormal marrow eosinophils
9872 = Acute myeloid leukemia, minimal differentiation
9873 = Acute myeloid leukemia without maturation
9874 = Acute myeloid leukemia with maturation
9875 = Atypical chronic myeloid leukemia BCR/ABL negative
9876 = Acute monocytic leukemia
9877 = Acute myeloid leukemia with multilineage dysplasia
9878 = Acute myeloid leukemia, t(8;21)(q22;q22)
9879 = Acute myeloid leukemia, 11q23 abnormalities
9880 = Acute megakaryoblastic leukemia
9881 = Therapy-related acute myeloid leukemia, NOS
9882 = Myelodysplastic syndrome, NOS
9883 = Refractory anemia with excess blasts
9884 = Refractory anemia with excess blasts in transformation
9885 = Refractory cytopenia with multilineage dysplasia
9886 = Myelodysplastic syndrome with 5q deletion (5q–) syndrome
9887 = Therapy-related myelodysplastic syndrome, NOS
9888 = Myelodysplastic syndrome, NOS
9889 = Myelodysplastic syndrome, NOS

* Usually considered of uncertain/borderline behavior

SUMMARY STAGE

1 Localized (isolated/mono-ostotic/single/solitary/unifocal for M-9731/3, 9734/3, 9750/3, 9751/3, 9752/3)
2 Distant (polyostotic); disease disseminated at diagnosis
3 Death certificate only case

Unknown

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.