Changes to the Hematopoietic and Lymphoid Neoplasm Coding Manual
KCR 2018 SPRING TRAINING

2018 Hematopoietic Database Updates

Updates were done to the Hematopoietic Database based on the WHO Hematopoietic and Lymphoid Neoplasms, Revised 4th edition, Volume II, published in 2017. Changes include:

• Revised preferred terminology for ICD-O-3 codes
• Default primary sites updated (9724, 9727, 9811-9818, 9827, 9837)
• Module information updated - Module 4 is for lymphoma/leukemia histologies; it is effective for cases diagnosed 2010-2017 only. For cases diagnosed 1/1/2018 forward, see the Hematopoietic database.

Note: Due to changes between 2010-2017 and 2018, it is very important to choose the diagnosis year when looking for information on a histology.
Primary site for cases diagnosed 01/01/2018 and forward

Assign primary site **C421 (Bone marrow)** when the histology is:
- 9724/3-Systemic EBV-positive T-cell lymphoma of childhood
- 9727/3-Blastic plasmacytoid dendritic cell neoplasm
  - 9811/3-B-lymphoblastic leukemia/lymphoma, not otherwise specified (NOS)
  - 9812/3-B-lymphoblastic leukemia/lymphoma with t(9;22)(q34.1;q11.2); BCR-ABL1
  - 9813/3-B-lymphoblastic leukemia/lymphoma with t(v;11q23.3); KMT2A-rearranged
  - 9814/3-B-lymphoblastic leukemia/lymphoma with t(12;21) (p13.2;q22.1); ETV6-RUNX1
  - 9815/3-B-lymphoblastic leukemia/lymphoma with hyperdiploidy
  - 9816/3-B-lymphoblastic leukemia/lymphoma with hypodiploidy (Hypodiploid ALL)
  - 9817/3-B-lymphoblastic leukemia/lymphoma with t(5;14)(q31.1;q32.1); IGH/IL3-
  - 9818/3-B-lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); TCF3-PBX1
  - 9837/3-T lymphoblastic leukemia/lymphoma

Primary site for cases diagnosed 01/01/2018 and forward

Assign primary site **according to lymphoma rules** when the histology is:
- 9827/3-Adult T-cell leukemia/lymphoma

In 2018 and forward, this will be considered a lymphoma. The primary site will be coded according to the rules in Modules 6 or 7, and not Module 4.
2018 Hematopoietic Database Updates

Other changes in the 2018 Hematopoietic Database:

- Updates to Immunophenotyping, Genetics information
- Update of transformations
- Grade field changed to “not applicable.”

New histology codes, (not approved for use in United States and Canada for 2018) will be coming in 2019.

<table>
<thead>
<tr>
<th>Change</th>
<th>New code</th>
<th>Term</th>
<th>Current code</th>
<th>Current Term</th>
</tr>
</thead>
<tbody>
<tr>
<td>Behavior</td>
<td>9709/1</td>
<td>Primary cutaneous CD4+ sm/medium T cell lymphoproliferative disorder</td>
<td>9709/3</td>
<td>Primary cutaneous T cell lymphoma</td>
</tr>
<tr>
<td>Behavior</td>
<td>9725/1</td>
<td>Hydroa vacciniforme-like lymphoproliferative disorder</td>
<td>9725/3</td>
<td>Hydroa vacciniforme-like lymphoma</td>
</tr>
<tr>
<td>Behavior</td>
<td>9751/1</td>
<td>Langerhans cell histiocytosis</td>
<td>9751/3</td>
<td>Langerhans cell histiocytosis</td>
</tr>
<tr>
<td>Behavior</td>
<td>9971/1</td>
<td>Polymorphic post-transplant lymphoproliferative disorder</td>
<td>9971/3</td>
<td>Polymorphic post-transplant lymphoproliferative disorder</td>
</tr>
</tbody>
</table>
New histology codes not yet approved for 2018 (coming in 2019)

<table>
<thead>
<tr>
<th>Change</th>
<th>Code</th>
<th>Term</th>
<th>Current</th>
<th>Term</th>
</tr>
</thead>
<tbody>
<tr>
<td>New</td>
<td>9715/3</td>
<td>Anaplastic large cell lymphoma, ALK neg</td>
<td>9702/3</td>
<td>Peripheral T cell lymphoma</td>
</tr>
<tr>
<td>New</td>
<td>9749/3</td>
<td>Erdheim-Chester disease</td>
<td>N/A</td>
<td>N/A</td>
</tr>
<tr>
<td>New</td>
<td>9766/3</td>
<td>Lymphomatoid granulomatosis, grade 3</td>
<td>9680/3</td>
<td>Diffuse large B cell lymphoma</td>
</tr>
<tr>
<td>New</td>
<td>9877/3</td>
<td>AML with gene mutations</td>
<td>9861/3</td>
<td>AML, NOS</td>
</tr>
<tr>
<td>New</td>
<td>9878/3</td>
<td>AML with biallelic mutation of CEBPA</td>
<td>9861/3</td>
<td>AML, NOS</td>
</tr>
<tr>
<td>New</td>
<td>9879/3</td>
<td>AML with mutated RUNX1</td>
<td>9861/3</td>
<td>AML, NOS</td>
</tr>
<tr>
<td>New</td>
<td>9912/3</td>
<td>AML with BRC-ABL1</td>
<td>9861/3</td>
<td>AML, NOS</td>
</tr>
<tr>
<td>New</td>
<td>9968/3</td>
<td>Myeloid/lymphoid neoplasms w/PCM2-JAK2</td>
<td>9975/3</td>
<td>MDS/Myeloproliferative neoplasm, unclassifiable</td>
</tr>
<tr>
<td>New</td>
<td>9980/3</td>
<td>MDS with single lineage dysplasia</td>
<td>9991/3</td>
<td>Refractory neutropenia</td>
</tr>
<tr>
<td>New</td>
<td>9980/3</td>
<td>MDS with single lineage dysplasia</td>
<td>9992/3</td>
<td>Refractory thrombocytopenia</td>
</tr>
<tr>
<td>New</td>
<td>9993/3</td>
<td>MDS with ring sideroblasts</td>
<td>9985/3</td>
<td>MDS with multilineage dysplasia</td>
</tr>
</tbody>
</table>

Number of primaries

The most important thing to remember from these rules is:
◦ These rules are for cancer registrars and are not followed by physicians. Follow the rules stated in this manual and abstract the number of primaries based on the rules. This may, or may not, agree with what the physician determines.

Multiple Primaries Calculator
◦ The multiple primaries calculator (MPC) is to be used ONLY when the rules instruct you to do so. Use of the MPC without applying the rules first may result in a wrong number of primaries and/or the wrong histologies.
Two important changes to the MP rules

Abstract as 2 primaries when a nodal MALT lymphoma occurs before or after an extranodal MALT lymphoma. These are 2 distinct lymphomas, with nodal lymphomas coded to lymph nodes (C77.0-C77.9) and extranodal coded to any other site.

Solitary Plasmacytomas (9731/3, 9734/3) occurring after a diagnosis of plasma cell myeloma (or multiple myeloma -9732/3) are NOT new primaries. The presence of plasmacytomas after multiple myeloma is evidence of advanced disease.

Diagnostic confirmation

Diagnostic Confirmation Coding Instructions for Hematopoietic and Lymphoid Neoplasms (9590/3-9992/3)

- Code 1: Positive histology (includes positive cytology)
- Code 2 is rarely used for Hematopoietic and Lymphoid neoplasms
Steps for using the hematopoietic database and coding manual

1. Identify the working (preliminary) histology code(s)

2. Determine the number of primaries using the working histology code(s) with the MP rules in the manual
   ◦ Note 1: The M rule references in the Heme DB are to be used as a guide only. Start with rule M1, move through the rules in consecutive order and stop at the first rule that applies.
   ◦ Note 2: Use the Hematopoietic Multiple Primaries Calculator in the Heme DB only when instructed by the rules in the Hematopoietic Manual.

3. Verify or revise the working histology code(s) using the PH rules in the manual

4. Determine primary site

5. Determine the grade (use only for cases diagnosed 2010-2017. Grade no longer collected for cases diagnosed 2018 and forward).

Changes to Other therapy

Collect blood-thinners and/or anti-clotting agents for essential thrombocytemia (9962/3) ONLY

Previously, instructions stated that blood thinners and/or anti-clotting agents were also collected for the histologies listed below. This change is effective for cases diagnosed 1/1/2010 and forward; however, there is no requirement of changing cases already abstracted

◦ 9740/3 Mast cell sarcoma
◦ 9741/3 Systemic mastocytosis with an associated hematological neoplasm
◦ 9742/3 Mast cell leukemia
◦ 9875/3 Chronic myeloid leukemia BCR-ABL1-positive
◦ 9950/3 Polycythemia vera
◦ 9961/3 Primary myelofibrosis
◦ 9963/3 Chronic neutrophilic leukemia
◦ 9975/3 Myelodysplastic/myeloproliferative neoplasm, unclassifiable
Immunotherapy

Donor Leukocyte Infusions – Always abstract as immunotherapy even if not listed in treatment section of the Hematopoietic Database

The End

Thank you!
For questions, contact
Frances Ross
fer@kcr.uky.edu