


National Cancer Institute


Hematopoietic and Lymphoid Neoplasm Project



Introduction to Hematopoietic and Lymphoid Neoplasm Rules

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Hematopoietic Working Group

- SEER Program National Cancer Institute
- National Program of Cancer Registries Centers for Disease Control (NPCR CDC)
- American College of Surgeons Commission on Cancer (ACoS CoC)
- National Cancer Registrars Association (NCRA)
- North American Association of Central Cancer Registries (NAACCR)
- Canadian Cancer Registries (CCR)

Hematopathology and Hematology/Oncology Subject Matter Experts

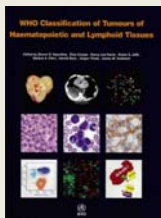
Why New Rules?

- 2009 Revised WHO Classification of Tumours of Hematopoietic and Lymphoid Tissues, 4th ed.
- Based on scientific and medical advances
- Address new terminology and new codes
- Address under-reporting of cases
- Need to update reporting rules
- Need to update coding rules
 - Single vs. Multiple Primary
 - Primary Site, Histology, Grade

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What is New / Different?

- Authoritative reference = WHO
- New histology terms
- New reportable conditions
- New Diagnostic Confirmation
- Transformation = New Primary



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Manual and Database

- Manual contains Case Reporting Rules, Coding Rules for primary site, histology, and grade, a detailed glossary, and a number of key tables
- Database contains detailed information on each condition, diagnostic testing, abstractor notes
- Manual and Database MUST be used together



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Manual	Database
<ul style="list-style-type: none"> • Reportable Instructions • Multiple Primary Rules • Primary Site Rules • Histology Coding Rules • Grade Coding Rules • Glossary • Tables 	<ul style="list-style-type: none"> • Neoplasm Definition • Neoplasm Synonyms • MP Calculator • Diagnostic Method • Genetic Testing • Immunophenotype • Treatment • Transformation • Abstractor Notes

New Histology Terms and Codes

New Histology Term	ICD-O Code
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Acute myeloid leukemia with inv(3)(q21q26.2) or t(3;3)(q21;q26.2); RPN1EVII	9869/3
Acute myeloid leukemia with t(6;9)(p23;q34) DEK-NUP214	9865/3
ALK positive large B-cell lymphoma	9737/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3

New Reportable Conditions

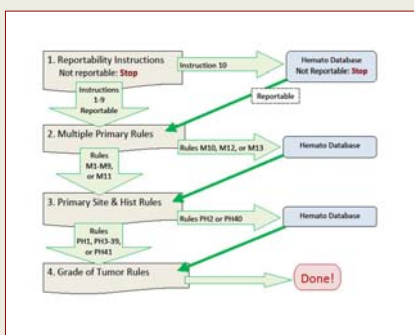
New Reportable Histology	ICD-O Code
Langerhans cell histiocytosis, NOS	9751/3
Myeloproliferative neoplasm, unclassifiable/Myelodysplastic/Myeloproliferative neoplasm, unclassifiable	9975/3
T-cell large granular lymphocytic leukemia/ Chronic lymphoproliferative disorder of NK-cells	9831/3

Using the Manual – 4 Questions

1. Is the condition reportable?
2. How many primaries do I abstract?
3. How do I code the primary site and histology?
4. How do I code the grade?

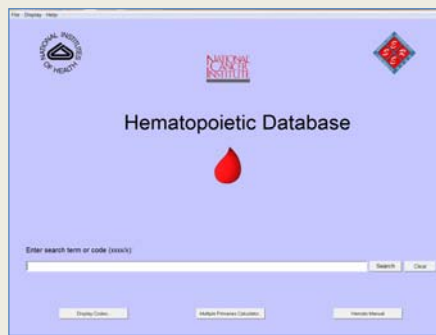
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Moving Through the Rules



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The Hematopoietic Database



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The Hematopoietic Database

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The Hematopoietic Database

Plasma cell myeloma

ICD-9.3 Code: Preferred Term
97303 Plasma cell myeloma

Alternate Names
Alpha PCM
Alpha plasma cell myeloma
Evolving multiple myeloma
Purpure plasma cell myeloma

Definitions
Multiple myeloma is a type of cancer of the plasma cells which are immune cells in bone marrow that produce antibodies.
Bone marrow-based, multifocal plasma cell neoplasm characterized by serum monoclonal protein, skeletal destruction with osteolytic lesions, and hypercalcemia.

Definitive Diagnostic Methods
FCM, Genetics, Immunophenotyping, Peripheral blood, Bone marrow aspiration, Bone Marrow biopsy, Bence-Jones protein

Disease Genetics Data
High load IGHV gene somatic hypermutation. Five major oncogenes involved in 14q32 translocation: cyclin D1, C-MYC, FGFR3/MGSET, cyclin D3, and MAFK. High load of IGHV gene somatic hypermutation. Whole or partial chromosome deletions or translocations, Trisomies, Immunoglobulin heavy and light chain genes are also affected.

Disease Immunophenotyping
MDSB1, CD138, CD19, CD38, CD79a, CD56 aberrantly expressed

Treatments
For more treatment information, see 112230

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The Hematopoietic Database

Abstractor Notes

Generalized bone marrow involvement is typically present. Lytic bone lesions and bone tumor masses of plasma cells also occur. Extramedullary involvement is generally a manifestation of advanced disease. In most patients there is a constellation of clinical, laboratory, radiographic, and pathologic findings. The patient often presents with end organ damage and bone pain. Bence-Jones protein accumulates in the renal tubules causing renal damage.

There are four clinical variants of plasma cell myeloma, all of which are coded to 97303. Those variants are: Asymptomatic (smoldering) plasma cell myeloma, non-secretory myeloma, plasma cell leukemia, and plasma cell leukemia.

Asymptomatic (smoldering) plasma cell myeloma. The diagnostic requirements for myeloma are met but there is no related organ or tissue impairment. Asymptomatic PCM is similar to MGUS in its lack of manifestations, but much more likely to progress to symptomatic plasma cell myeloma. About 6% of patients with myeloma are initially asymptomatic.

Inactive or smoldering plasma cell myeloma is asymptomatic. The bone marrow is involved but there is no related organ or tissue impairment. Patients may have stable disease for long periods of time but the cumulative probability of progression to symptomatic plasma cell myeloma or amyloidosis increases over the years.

Non-secretory myeloma. In approximately 2% of plasma cell myeloma there is an absence of an M protein on immunofixation electrophoresis. Cytoplasmic M protein is present in the neoplastic plasma cells in about 95% of these when evaluated by immunohistochemistry, consistent with impaired secretion of Ig.

Plasma cell leukemia (PCL). The number of plasma cells in the peripheral blood exceeds 2000/sqmm (5% of the leukocyte differential count). In addition to the peripheral blood and bone marrow, the neoplastic plasma cells may be found in extramedullary tissue such as spleen, liver, pleural effusions, ascites, and cerebrospinal fluid. PCL may be present at diagnosis (primary PCL) or occur as a late feature of plasma cell myeloma (secondary PCL). Typically the immunophenotype of PCL differs from other myelomas by the lack of aberrant CD56 expression. Most clinical features are similar to plasma cell myeloma but osteolytic lesions and bone pain are less frequent and lymphadenopathy, organomegaly, and renal failure are present more often. PCL is an aggressive disease with short survival.

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Heme/Lymph Training Modules

- Background / Introduction
- Disease Presentation and Diagnostic Process
- Hematopoietic Cell Lineages – Myeloid/Lymphoid
- Instructions for Assessing Case Reportability
- Coding Rules – Primary Site, Histology, Grade
- How to Use the Manual and the Database
- Using the Tables in the Manual Correctly
- <http://www.seer.cancer.gov/tools/heme/training>

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Summary – Heme/Lymph Rules

- Reference: WHO Preferred Terms and Codes
- Use Manual and Database as Companion Tools
- Cases diagnosed on or after January 1, 2010.

- Registrars **MUST** download the Hemato Database
- Registrars **SHOULD** view the 13 Training Modules
- GO TO SEER <http://seer.cancer.gov/tools/heme>

- Email NCISEERQI@mail.nih.gov with questions

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