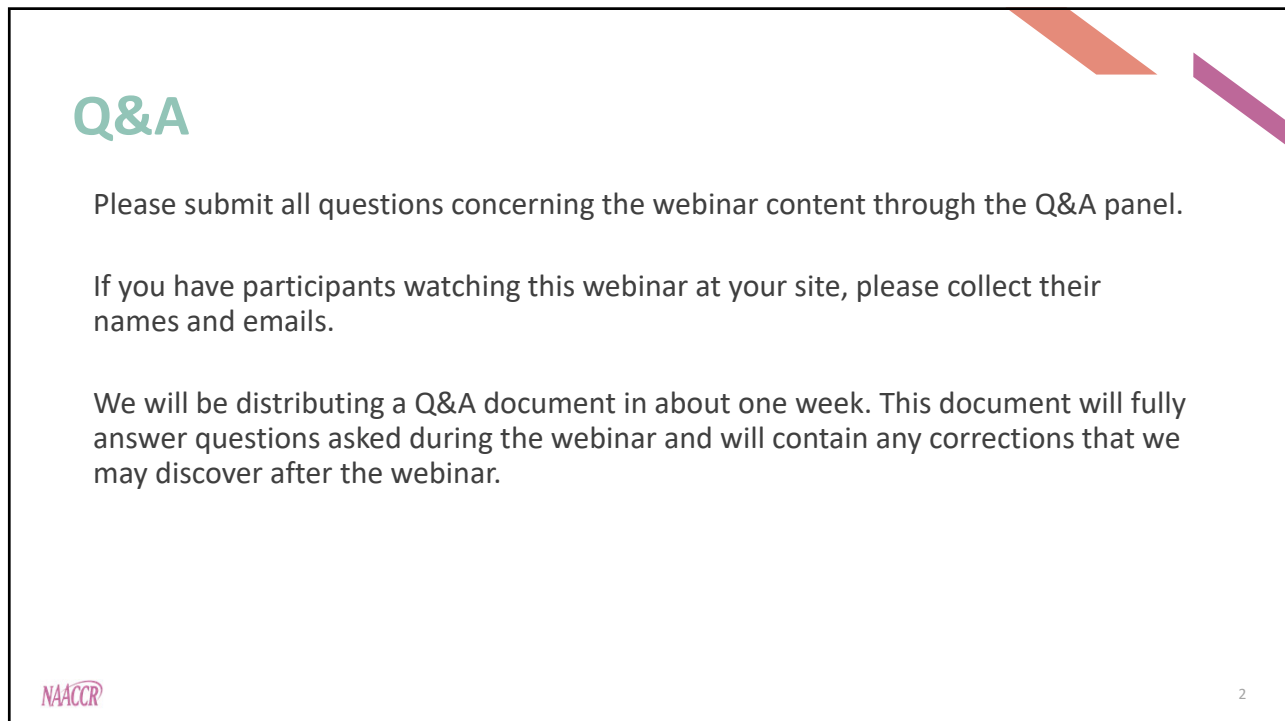




1



2

## Fabulous Prizes

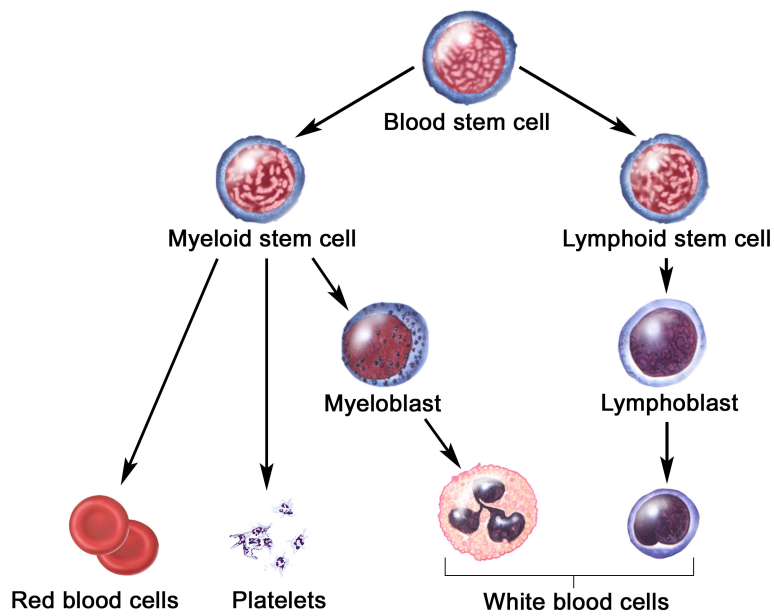


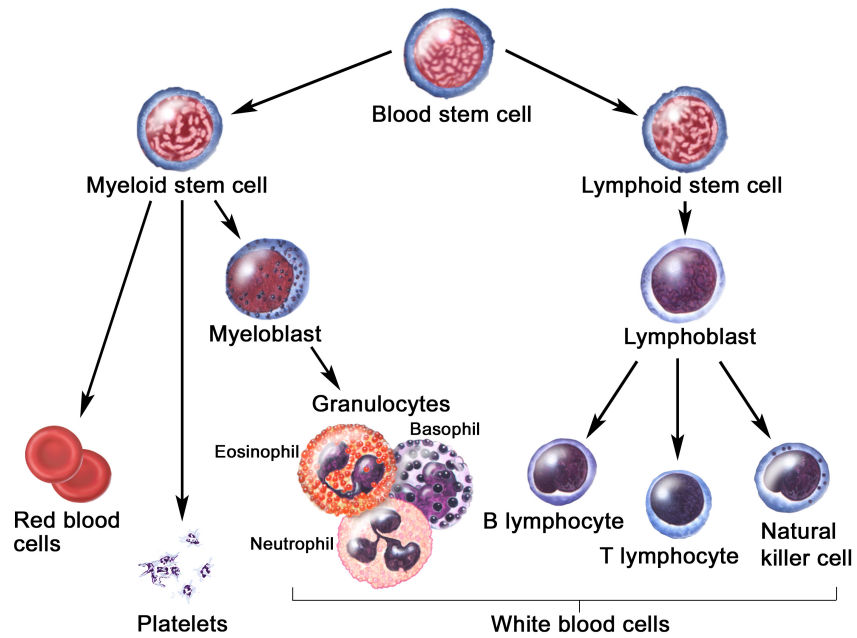
## Guest Presenter

- Denise Harrison, CTR
  - 2021-2022 Board of Directors, NCRA
- Louanne Currence, RHIT, CTR
  - Cancer Registrar, North Kansas City Hospital

# Abstracting Hematopoietic Neoplasms

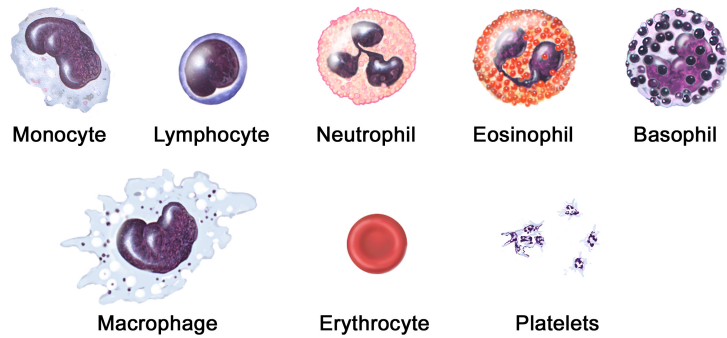
Denise Harrison, BS, CTR  
Louanne Currence, CTR





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### Blood Cells



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## Common Types of Leukemia

### Adult

- Acute myeloid (myelogenous)/AML
- Chronic myeloid (myelogenous)/CML
- Acute lymphocytic (lymphoblastic)/ALL
- Chronic lymphocytic/CLL

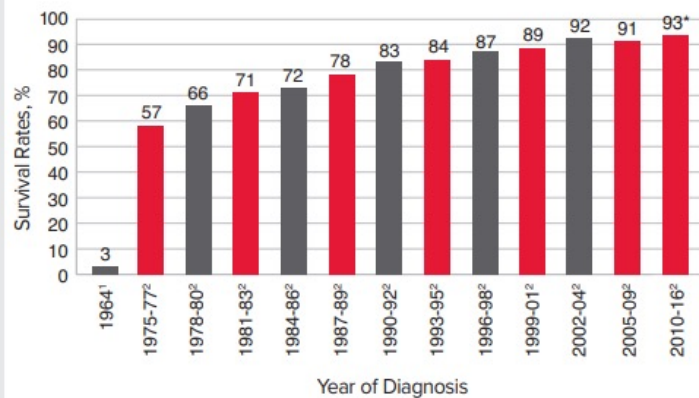
### Childhood

- Acute lymphocytic (lymphoblastic)/ALL
- Acute myelogenous (myeloid) AML
- Hybrid or mixed lineage
  
- Chronic leukemias are RARE in children
- Chronic myelogenous/CML
- Chronic lymphocytic/CLL

5

## ALL Success Story

**Five-Year Relative Survival Rates for Acute Lymphoblastic Leukemia in Children Under 15, Diagnosed 1964-2016**



**Figure 9.** Sources: 1. Zuelzer WW. Implications of long-term survivals in acute stem cell leukemia of childhood treated with composite cyclic therapy. *Blood*. 1964;24:477-494. 2. *SEER (Surveillance, Epidemiology, and End Results) Cancer Statistics Review, 1975-2017*. National Cancer Institute; 2020.

\* The difference in rates between 1975-1977 and 2010-2016 is statistically significant ( $P < .05$ ).



## Children are not Short Adults

---

- NCI-funded study in 2018 found AML is a greatly different disease in children than adults
- TARGET study included ALL, AML, neuroblastoma, Wilms tumor and osteosarcoma
  - Research identified 142 altered genes, 45% found in adult cancer

7



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## Diagnostic Confirmation

8

# Diagnostic Process

## Leukemia

- Patient presents w/ unexplained weight loss, weakness, chronic fatigue, and/or easy bruising which prompts workup
- CBC/peripheral blood smear: if abnormal, then
  - Bone Marrow biopsy
    - Immunophenotype/genetic information generally required to identify a specific histology

## Lymphoma

- Biopsy of lymph node, organ, or mass
- Careful! Do not assume the biopsy site is the primary site
  - Most accessible site is the one biopsied
- Hodgkin Lymphoma characterized by Reed-Sternberg cells

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# Diagnostic Confirmation Codes

Microscopically Confirmed		NOT Microscopically Confirmed	
1	Positive histology • Includes: peripheral blood smear only	5	Positive laboratory test/marker study • Includes cases with positive immunophenotyping or genetic studies and no histological confirmation • Does NOT include cases where a peripheral blood smear is done (code 1) or peripheral blood smear followed by flow cytometry (code 3)
2	Positive cytology	6	Direct visualization w/out microscopic confirmation
3	Positive histology PLUS: • Positive immunophenotyping AND/OR • Positive genetic studies • Includes peripheral blood smear followed by flow cytometry	7	Radiology and other imaging techniques w/out microscopic confirmation
4	Positive microscopic confirmation, method not specified	8	Clinical diagnosis only (other than 5, 6, or 7)

9 Unknown whether or not microscopically confirmed; death certificate only

10



## Diagnostic Confirmation Codes 1 and 3

Use code 1 (**Positive** histology) when the diagnosis is based solely on tissue: Bone marrow, LN, organ, peripheral blood smear; (CBC and WBC – Leukemia only)

- Positive histology and
  - Immunophenotyping, genetic testing, or JAK2
    - NOT done
    - Done, but negative for disease being abstracted
    - ~~Done, but not listed in Definitive Dx Methods~~ (See new note 1 next slide)
  - IHC studies done, but provisional NOS dx or 1 or more provisional dx
  - Historical cases not already in DB when info states there was histo confirmation

Use Code 3 **Positive** histology (Code 1) PLUS **positive** immunophenotyping or genetic testing when 2010+ dx AND:

- Positive histology AND
  - Immunophenotyping, genetic testing, or JAK-2 testing listed in Definitive Dx Methods
    - Confirms diagnosis OR
    - Identifies more specific histology
    - NOS histology dx'd (not a provisional dx)
- Do not use Code 3 when test:
  - Identifies more specific histology but uses ambiguous terminology OR
  - Result preceded by “patchy weak staining”

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## Diagnostic Confirmation – New Note 1

- Hematopoietic manual: Coding Diagnostic Confirmation, Code 3 (pg. 19)
  - New Note 1: While every attempt is made to keep the Hematopoietic database updated, it is impossible to keep the Hematopoietic database updated with all the immunophenotyping or genetics that can be done for a specific histology since clinical medicine continues to evolve. If immunophenotyping or genetics are used by the pathologist/managing physician to identify a specific neoplasm that are not included in the Hematopoietic database, and Genetic testing and/or Immunophenotyping are listed as Definitive Diagnostic Methods for that histology, go ahead and use these.

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## Diagnostic Confirmation

- Review “Definitive Diagnostics Methods” section in the Hematopoietic Database
  - If genetics or immunophenotyping are
    - Listed, the histology can have a diagnostic confirmation of 3
      - If the pathology report lists
        - One of these that confirms the diagnosis, code 3
        - Other genetics or immunophenotyping that confirms the diagnosis but is not listed in the Hematopoietic database, assign 3
          - » As noted in previous slide, it is very difficult to keep up with all the different tests used
    - Not listed, the histology cannot have a diagnostic confirmation of 3

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## Diagnostic Confirmation – New Note 2

Note 2: These are histologies that are defined by positive genetics and/or immunophenotyping or genetics and **must always have diagnostic confirmation code 3** (Edits enforced 2022+)

Genetics and/or immunophenotyping	Genetics Only
<b>Table B5: Myelodysplastic Syndromes</b>	<b>Table B6: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms</b>
9986/3	9865/3
<b>Table B7: Acute Leukemias of Ambiguous Lineage</b>	9866/3
9806/3	9869/3
9807/3	9871/3
9808/3	9877/3
9809/3	9878/3
<b>Table B8: Precursor Lymphoid Neoplasms</b>	9879/3
9812/3	9896/3
9813/3	9897/3
9814/3	9911/3
9815/3	9912/3
9816/3	9965/3
9817/3	9966/3
9818/3	9967/3
	9968/3

All histologies listed (both columns) are primarily diagnosed based on bone marrow biopsy/peripheral blood; however, LN and/or organ tissue biopsies may also be used for those listed under Table B8.

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## Diagnostic Confirmation Must be 3

---

### New Diagnostic Confirmation section in Heme Database

- This AML is part of the "AML with recurrent genetic abnormalities" group. Since this AML is diagnosed based on genetics, diagnostic confirmation will always be 3.
- This histology can only be determined by positive genetics and/or immunophenotyping, diagnostic confirmation will always be 3.

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## Example: Diagnostic Confirmation Must be 3

---

### 9986/3 Myelodysplastic syndrome with isolated del(5q)

#### Definitive Diagnostic Methods

Bone marrow biopsy  
Genetic testing

#### Genetics Data

Deletion between bands q31 and q33 on chromosome 5

#### Immunophenotyping

None

#### Diagnostic Confirmation

This histology can only be determined by positive genetics and/or immunophenotyping, diagnostic confirmation will **always** be 3.

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## New Diagnostic Confirmation Field in Heme DB Diagnostic Confirmation Must be 3

9912/3: [Acute](#) myeloid [leukemia](#) with BCR-ABL1

Help me code for diagnosis year : ←

2021 ←

Coding Manual: [Hematopoietic Coding Manual \(PDF\)](#)

### Abstractor Notes

(This [code](#) is effective for [cases](#) diagnosed 2021 and later. For [cases](#) diagnosed prior to 2021 see [code](#): 9861/3.)

Patients most commonly present with [leukocytes](#) with a blast predominance and [variable](#) presence of [anemia](#) and [thrombocytopenia](#).

This is an [aggressive](#) disease, with poor [response](#) to traditional AML [therapy](#) or [tyrosine kinase inhibitor therapy](#) alone. Improved [survival](#) is seen with [tyrosine kinase inhibitor](#) followed by [allogeneic hematopoietic cell transplantation](#).

### Diagnostic Confirmation

This AML is part of the "AML with recurrent genetic abnormalities" group. Since this AML is diagnosed based on genetics, diagnostic confirmation will always be 3.

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## Example: Diagnostic Confirmation Must be 3

9815/3 B-lymphoblastic leukemia/lymphoma with hyperdiploidy

### Definitive Diagnostic Methods

Bone marrow biopsy  
FISH  
Genetic testing  
Immunophenotyping  
Karyotyping

### Genetics Data

Extra copies of [chromosomes](#) 21, X, 14 and 4

### Immunophenotyping

CD10+  
CD19+  
CD34+  
CD45 absent

### Diagnostic Confirmation

This histology can only be determined by positive genetics and/or immunophenotyping, diagnostic confirmation will **always** be 3.

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## Diagnostic Confirmation Cannot be 3

- New Note 3: The following histologies should never be assigned diagnostic confirmation 3 since they are nonspecific codes and neither genetic testing or immunophenotyping are listed as Definitive Diagnostic Methods for these histologies. If there is immunophenotyping or genetics available, then a more specific histology code may be able to be assigned (Edits enforced 2022+)

No Genetics or Immunophenotyping Listed				
9590/3	9800/3	9860/3	9980/3	9989/3
9655/3	9820/3	9863/3	9982/3	9991/3

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## Diagnostic Confirmation – New Note 3

- These are histologies that are never assigned diagnostic confirmation code 3 (edits enforced 2022+) because they are non-specific codes and neither genetic testing nor immunophenotyping are listed as Definitive Diagnostic Methods

9590/3	9863/3	If genetics and/or immunophenotyping are available, re-review to see if a more specific neoplasm can be coded
9655/3	9980/3	
9800/3	9982/3	
9820/3	9989/3	
9860/3 (obs)	9991/3*	

\* Code used for 2010-2020 dx; assign 9980 for 2021+

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## Example: Diagnostic Confirmation Cannot be 3

### 9590/3 Malignant lymphoma, NOS

#### Definitive Diagnostic Methods

Clinical diagnosis  
Histologic confirmation

#### Genetics Data

None

#### Immunophenotyping

None

No Genetics or Immunophenotyping listed; therefore, histology **cannot** have a diagnostic confirmation code of 3

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## New Diagnostic Confirmation Field in Heme DB Diagnostic Confirmation Cannot be 3

9590/3: [Malignant lymphoma](#), NOS

Help me code for diagnosis year :

2021

Coding Manual: Hematopoietic Coding Manual (PDF)

#### Abstractor Notes

This NOS [histology](#) is a [generic](#) disease description. [DCO cases](#) or path report only [cases](#) may stay in this classification. In most [cases](#), an NOS [histology](#) is only the working [diagnosis](#); the physician will run further [diagnostic procedures](#) and look for various clinical presentations to identify a more specific disease.

Further review of the [medical record](#) should be performed to look for the tests listed as [definitive diagnosis](#). When a more specific [diagnosis](#) is identified, the [histology](#) should be changed to the more specific neoplasm name and [code](#).

For [9590/3 malignant lymphoma, NOS](#), [non-Hodgkin lymphoma](#), classical [Hodgkin lymphoma](#), and any specific Hodgkin and non-Hodgkin lymphomas would be a more specific [histology](#).

See the [histology tables](#) ([Appendix B](#) of the [Hematopoietic manual](#)) for more information on NOS and more specific histologies.

#### Diagnostic Confirmation

This is a histology for which the Definitive Diagnostic Method does not include Genetics Data or Immunophenotyping, thus Diagnostic Confirmation should never be 3. If genetics and/or immunophenotyping are available, re-review to see if a more specific neoplasm can be coded.

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## Diagnostic Confirmation Can be 3

### New Diagnostic Confirmation section in Heme Database

- This histology can be determined by positive histology (including peripheral blood) with or without genetics and/or immunophenotyping. Review the Definitive Diagnostic Methods, Immunophenotyping and Genetics Data sections below, and the instructions in the Hematopoietic Manual for further guidance on assigning Diagnostic confirmation.

This applies to the **majority** of histologies in the heme db.

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## New Diagnostic Confirmation Field in Heme DB Diagnostic Confirmation Can be 3

9985/3: [Myelodysplastic syndrome](#) with multilineage [dysplasia](#)

Help me code for diagnosis year :

2021

Coding Manual: [Hematopoietic Coding Manual \(PDF\)](#)

### Abstractor Notes

**Blood** and **bone marrow** are always **involved**. At least 2 types of **blood** counts are low and have an **abnormal** appearance under the microscope (**dysplasia**). The number of blasts is less than 5%.

This **histology code** also includes childhood MDS. MDS is very rare in children. Both the **peripheral blood** and **bone marrow** are **involved**.

For MDS diseases (9980, 9982, 9983, **9985**, 9986, 9989, 9991, 9992, 9993), **abstracting** each of the subtypes would result in over-counting of the diseases.

1. **Code** only the first subtype that is diagnosed.

2. Do not change the **histology code** or create a new **abstract** for any subsequent specific MDS subtypes.

### Diagnostic Confirmation

This histology can be determined by positive histology (including peripheral blood) with or without genetics and/or immunophenotyping. Review the Definitive Diagnostic Methods, Immunophenotyping and Genetics Data sections below, and the instructions in the Hematopoietic Manual for further guidance on assigning Diagnostic confirmation.

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## Diagnostic Confirmation

---

- Per Diagnostic Confirmation, Code 1, #3
  - Peripheral blood smear
    - Can be used as a histological diagnosis for any of the hematopoietic histologies (9590/3-9992/3)
    - NOTE: STORE 2022 page 137 (for code 1) states peripheral blood smear can be used only for leukemia (not other heme histologies)
- Per recent clarification:
  - Peripheral blood smear followed by flow cytometry (most commonly done with CLL/SLL [9823/3]) is Diagnostic confirmation code #3 (See Heme Manual, Code 3: 1c)
    - If unable to find documentation that a peripheral blood smear was done first, assume that it was and code 3
    - The flow cytometry is what is confirming the diagnosis

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---

## First Course of Treatment for Hematopoietic Neoplasms

26



## What is Treatment?

### Cancer directed treatments

- Surgery
- Radiation
- Chemotherapy
- Immunotherapy
- Hormone therapy
- Bone marrow transplant

### Non-Cancer-directed (passive) treatments

- Observation (active surveillance), supportive care, or another type of treatment that does not meet the usual definition of treatment that “modifies, controls, removes or destroys proliferating cancer tissue.”

For the purposes of determining MPs (Rules M10-M13) in heme diseases, “treatment” refers to cancer-directed treatment, rather than **passive** treatments such as **supportive care or observation**

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## Treatment Notes

- Code tx on both abstracts when patient has multiple primaries and tx given would affect both
- EX: Patient dx'd 5/2018 w/ multiple myeloma and mantle cell lymphoma (separate primaries per M15: Starts Velcade for myeloma.

SEER Rx Interactive Antineoplastic Search

Search Text:

Result

[D] - Bortezomib

### Primary Site

[Breast](#)

[colorectal](#)

[leukemia](#)

[lung cancer](#)

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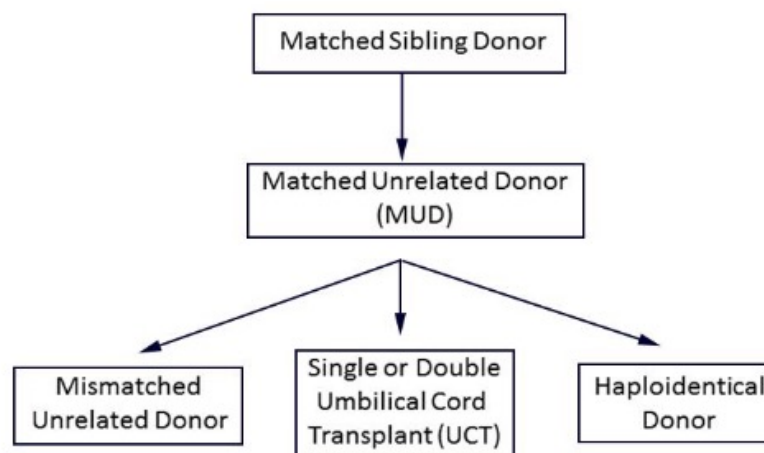
## Phlebotomy, Blood-Thinners/or Anticoagulants, & Transfusions

Effective with 2010 diagnoses and forward

- Do NOT collect blood transfusions as treatment
- Collect
  - Phlebotomy for polycythemia vera ONLY
  - Blood thinners, anticoagulants and/or anti-clotting agents for essential thrombocythemia ONLY
- Formerly 8 additional heme diseases on this list but tx protocols have changed
- No requirement we to recode old cases

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## Allogenic Stem Cell Transplant



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## Allogeneic (Someone Else's!)

---

### Reportable conditions

- ALL, AML
- CLL, CML
- Hodgkin lymphoma
- Multiple myeloma
- MDS
- MPD
- Non-Hodgkin lymphoma

### Non-reportable conditions

- Anemias
- Epidermolysis bullosa
- Hemophagocytic lymphohistiocytosis
- Metabolism errors
- Paroxysmal nocturnal hemoglobinuria
- SCID
- Thalassemia major
- Others

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## Autologous (Mine!)

---

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## Donor Leukocyte Infusions

---

- Donor leukocyte infusion (AKA buffy coat, DLI)
  - Infusion of lymphocytes, specifically T-cells, from the transplant donor
  - Use of DLI increasing, particularly for leukemia
  - Code as immunotherapy, even when not listed in treatment section of Heme DB
  - Document the DLI in the immunotherapy text field along with graft type, when available
    - No text field available for transplant/endocrine procedure

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---

## Case Reportability

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## Case Reportability – 7 Instructions

1. Search Heme DB
2. Report 9590-9992
  - Behavior of /3
  - Behavior of /1 as /3 when described as malignant by physician
3. Do not report in situ (/2) lymphomas

### Name

Myelodysplastic syndrome, unclassifiable

### ICD-O-2 Morphology

9989/1: Myelodysplastic syndrome, NOS

### ICD-O-3 Morphology Effective 2001 and later

9989/3: Myelodysplastic syndrome, unclassifiable

### Reportable

for cases diagnosed 2001 and later

### Primary Site(s)

C421

Primary site must be bone marrow (C421)

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## In Situ Lymphomas (1/1/2021+ Diagnoses)

- Case Reportability Instruction #3 in Heme Manual
  - Do NOT report in situ (/2) lymphomas;
    - Behavior is /1, even with the in situ diagnosis
    - /2 applies only to solid tumors
- 9673/1: In situ mantle cell neoplasia
  - In situ mantle cell lymphoma, ISMCN, Mantle cell lymphoma-like B cells of uncertain/undetermined significance
- 9695/1: In situ follicular neoplasia
  - Follicular lymphoma in situ, In situ follicular lymphoma, Intrafollicular neoplasia, ISFN

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## Case Reportability, cont.

---

4. Ambiguous terms
  - Report dx when:
    - Dx preceded by a listed ambiguous term (other than cytology or tumor markers)
      - Equivalent terms may be used [ex: favored instead of favor(s)]
    - Reportable and non reportable term both appear in medical record
  - **Do not report cases:**
    - Dx'd only by ambiguous cytology or tumor markers
    - If bx or MD statement confirms a non-reportable condition or disproves the ambiguous dx
  - Do not substitute synonyms of ambiguous terms (ex: supposed for presumed) or “likely” for “most likely”

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## Case Reportability Instructions

---

5. Report the case when the patient is treated for a reportable neoplasm.
  - Use NCI's Physicians' Data Query (PDQ) website at <http://www.cancer.gov/cancertopics/pdq> or the SEER\*Rx Antineoplastic Drugs Database
6. Report the case when there is a clinical diagnosis (physician's statement) of reportable hematopoietic or lymphoid neoplasm.
7. Report the case when a reportable diagnosis appears in any text or report described as a Definitive Diagnostic Method in the Heme DB.

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## ICDO-3.2 Effective 1/1/2021 Change of Behavior from /3 to /1

Code	Term
9702/1	Indolent T-cell lymphoproliferative disorder of the GI Tract
9709/1	Primary cutaneous CD4-positive small/medium T-cell lymphoma (C44. _)
9725/1	Hydroa vacciniforme-like lymphoma
9751/1	Langerhans cell histiocytosis, NOS
9751/1	Langerhans cell histiocytosis, monostotic
9751/1	Langerhans cell histiocytosis, polyostotic
9971/1	Polymorphic post transplant lymphoproliferative disorder

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## ICDO-3.2 Effective 1/1/2021- Change of Code

From	To	Term
9680/3	9766/3	Lymphomatoid granulomatosis, grade 3
9702/3	9715/3	Anaplastic large cell lymphoma, ALK negative
9811/3	9819/3	B lymphoblastic leukemia/lymphoma, BCR-ABL1-like
9826/3	9687/3	Acute leukemia, Burkitt type Acute lymphoblastic leukemia, mature B-cell type
		B-ALL Burkitt cell leukemia FAB L3
9861/3	9877/3	Acute myeloid leukemia with mutated NPM1
	9878/3	Acute myeloid leukemia with biallelic mutation of CEBPA
	9879/3	Acute myeloid leukemia with mutated RUNX1
	9912/3	Acute myeloid leukemia with BCR-ABL1
9975/3	9968/3	Myeloid and lymphoid neoplasm with PCM1-JAK2
9985/3	9993/3	Myelodysplastic syndrome w/ ring sideroblasts and multilineage dysplasia
9991/3	9980/3	Refractory neutropenia
9992/3	9980/3	Refractory thrombocytopenia

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## ICDO-3.2 Effective 1/1/2021 New Code and Term

---

Code	Term
9680 /1	EBV-positive mucocutaneous ulcer
9738 /1	HHV8-positive germinotropic lymphoproliferative disorder
9823 /1	Monoclonal B-cell lymphocytosis, CLL-type
9749 /3	Erdheim-Chester disease

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## Multiple Primary Rules

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## Multiple Primary (M) Rules

### General Instructions

---

1. Start with M1 and stop at the **first** rule that applies
  - Use the M rule references in the Heme DB “as a guide only”
  - M rules M8-M13 send us straight to M14 (pre-2021 dx) or M15 (2021+ dx) when they do not apply (not a transformation)
2. Chronic neoplasms have the potential to transform into another more acute neoplasm
  - Acute and chronic neoplasms listed in Transformations to and from section of **Heme DB**
3. Physician may start w/provisional dx(s), and move to a more specific dx as testing is completed
  - Provisional diagnoses are NOT multiple primaries
4. Use Multiple Primaries calculator ONLY when the rules instruct you to do so (Rules M4, M7, and M15)

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## M Rules

### SP Single Primary – MP Multiple Primaries

---

- M1** Minimal info available (DCO or path report only case) – **SP**
- M2** Single histology – **SP**
- Bilateral LNs &/or organs w/ single histology
    - Example: Rt and Lt breast w/ DLBCL
  - Recurrence of same histology – No timing rules
  - **EXCEPTION:** MALT lymphomas 9699/3 of LN (C77.x) before or after extranodal MALT 9699/3 – **MP**
- Example**
- 2013: MALT of Rt inguinal node, stage I, no recurrence.
  - 2018: ocular MALT; Stage III (New primary)

**MALT = mucosa-associated lymphoid tissue**

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## M Rules

SP Single Primary – MP Multiple Primaries

---

**M3** Sarcoma dx'd simultaneously or after a leukemia of same cell line – **SP**

- Mast cell sarcoma and mast cell leukemia
- Myeloid sarcoma and AML or another leukemia of myeloid origin (9840/3, 9865/3-9867/3, 9869/3-9874/3, 9891/3, 9895/3-9898/3, 9910/3, 9911/3 and 9931/3)

▪ **EXCEPTION:** CML codes (9863/3, 9875/3, 9876/3) ≠ same lineage as myeloid sarcomas – **MP**

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## Same Anatomic Location

---

Definition of same anatomic location which is used in Rules M4 and M5 (on next slide):

**same anatomic location** = same LN, same LN region(s), same organ(s), same tissue(s)

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## M Rules

### SP Single Primary – MP Multiple Primaries

---

**M4** ≥ 2 types NHL simultaneously present in same anatomic location – **SP**

**EXCEPTION:** Do not use for cutaneous NHL (other than NOS and more specific: go straight to rule M15 (use the MPC)

**M5** Both Hodgkin and NHL simultaneously present in same anatomic location – **SP**

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## M Rules

### SP Single Primary – MP Multiple Primaries

---

**M6** Hodgkin lymphoma in one location and NHL in another location – **MP**

**M7** More specific histology dx'd after NOS histology only when MP calculator confirms both histo codes = same primary – **SP**

- More specific histo can be in a different anatomic site
- No time restrictions
- Change the histology on the original abstract (if original is in your DB) and use the heme DB or previous editions of ICD-O to assign the code applicable to the original year of dx

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## Rules M8-M13

- Apply only when there is a transformation:
  - From a chronic neoplasm TO an acute neoplasm OR
  - From an acute neoplasm TO a chronic neoplasm
- Look in the heme DB
  - “Transformations to” (acute neoplasms) and “Transformations from” (chronic neoplasms) are defined for each applicable histology
  - Skip rules M8-M13 and go to Rule M14 (if diagnosis is prior to 2021), or M15 (when the diagnosis is 2021+) when:
    - No “Transformation to” or “Transformation from” is listed
    - OR, the neoplasms in question are not listed as “Transformations to” or “Transformations from” each other

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## Distinguishing Acute vs. Chronic Neoplasms

### Follicular lymphoma

Transformations to  
[9680/3 Diffuse large B-cell lymphoma, NOS](#)  
Transformations from  
None

Follicular lymphoma transforms **to** DLBCL; therefore, FL is the chronic form and DLBCL is the acute form.

### Plasma cell myeloma

Transformations to  
None  
Transformations from  
[9731/3 Solitary plasmacytoma of bone](#)  
[9734/3 Extrasosseous plasmacytoma](#)

Plasma cell myeloma does not transform **to** anything; therefore, Plasma cell myeloma is the acute form and the 2 neoplasms under “Transformations from” are the chronic forms.

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# Distinguishing Acute vs. Chronic Neoplasms

## DLBCL

Transformations to

None

Transformations from

- [9651/3 Lymphocyte-rich classical Hodgkin lymphoma](#)
- [9653/3 Lymphocyte-depleted classical Hodgkin lymphoma](#)
- [9659/3 Nodular lymphocyte predominant Hodgkin lymphoma](#)
- [9670/3 Malignant lymphoma, small B lymphocytes, NOS](#)
- [9671/3 Lymphoplasmacytic lymphoma](#)
- [9675/3 Malignant lymphoma, mixed small and large cell, diffuse](#)
- [9688/3 T-cell/histiocyte-rich large B-cell lymphoma](#)
- [9689/3 Splenic marginal zone lymphoma](#)
- [9690/3 Follicular lymphoma](#)
- [9691/3 Follicular lymphoma, grade 2](#)
- [9695/3 Follicular lymphoma, grade 1](#)
- [9698/3 Follicular lymphoma, grade 3](#)
- [9699/3 Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue \(MALT lymphoma\)](#)
- [9761/3 Waldenstrom macroglobulinemia](#)
- [9762/3 Heavy chain diseases](#)
- [9823/3 Chronic lymphocytic leukemia/small lymphocytic lymphoma](#)
- [9940/3 Hairy cell leukemia](#)

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## Overview of Rules M8-M13 (The “Transformation” rules)

Chronic and Acute Diagnosed Simultaneously or ≤ 21 Days			
# + Bxs	New Primary	Rule	Comments
1	No	M8	Abstract the <b>acute</b> neoplasm
Unknown	No	M9	Abstract the <b>later</b> diagnosis
2*	Yes	M11	Exception: Plasmacytoma & Multiple myeloma = SP
Acute Diagnosed >21 Days After Chronic			
	New Primary	Rule	Comments
	Yes	M10	Only applies to multiple myeloma following plasmacytoma when <u>completed</u> workup reveals single plasmacytoma, then dx of multiple myeloma > 21 days later
Chronic Diagnosed >21 Days After Acute			
Tx	New Primary	Rule	Comments
No	No	M12	Ex: 3/16/18 DLBCL, no Tx; 4/18/18 FL
Yes	Yes	M13	Exception: Plasmacytoma(s) after Multiple myeloma

\* 1 bx confirms the chronic and 1 bx confirms the acute form of the neoplasm

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## M Rules

### SP Single Primary – MP Multiple Primaries

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- M14** For 2010-2020 dx: Post-transplant lymphoproliferative disorder (PTLD) diagnosed *simultaneously* with any B- or T-cell NHL or Hodgkin lymphoma or plasmacytoma/myeloma – **SP**
- This is a change from previous instructions which listed lymphomas as PTLT transformations
    - If lymphoma **AFTER** a PTLT dx'd 2010-2020 – **MP**
  - Effective 1/1/21, PTLT w/out an associated lymphoma or plasmacytoma is /1 and not reportable
    - If lymphoma **AFTER** a PTLT dx'd 2021+ – **SP** (the lymphoma)
- M15** Use Heme DB Multiple Primaries Calculator to decide **SP vs MP**

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# Primary Site and Histology Rules

## 9 Modules

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## Primary Site Coding Instructions

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1. Use Heme Manual instructions, PH Rules, and Heme DB to code primary site
2. Do not use C423 (reticuloendothelial system NOS) or C424 (hematopoietic system NOS) for heme neoplasms
3. Primary Site Coding Instructions: 2 fields
  - Primary site: when applicable, a specific site will be listed
  - Primary site text: info on common primary sites moved from Abstractor Notes

Example: “Primary site must be bone marrow” for leukemias

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## Database Help

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### CLL/SLL

Primary Site(s)

See Module 3: Rules PH5, PH6

Most common sites of involvement: bone marrow, peripheral blood, lymph nodes

### Waldenstrom Macroglobulinemia (9761/3)

Primary Site(s)

C421

Primary site must be bone marrow (C421) for cases diagnosed 1/1/2018 and forward. For cases diagnosed 2010-2017, primary site must be blood (C420).

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## Abstractor's Notes

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### Abstractor Notes

**Waldenstrom** Macroglobulinemia (WM) is a subset of lymphoplasmacytic lymphoma.

Patients with WM have IgM (immunoglobulin M) in their blood and/or bone marrow. IgM is also called IgM monoclonal gammopathy. There will also be an increased number of lymphocytes in the blood. A familial disposition may exist in up to 20% of patients. It tends to be diagnosed at a younger age than other lymphoplasmacytic lymphomas.

Treatment of **Waldenstrom** macroglobulinemia may include the following:

1. Biologic (immuno) therapy with interferon
2. Chemotherapy
3. Plasmapheresis
4. Proteasome inhibitor
5. Watchful waiting

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## Primary Site Coding Instructions, cont.

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### 4. Code primary site using (no hierarchy)

- Scans
- Medical record documentation
- Pathology report
- Heme DB

#### Notes:

- Do not simply code site of LN bx; use info from scans
- Path report not default for determining primary site, especially for lymphoma; standard depends on specific histology
- Assign bone marrow (C421) if dx by peripheral blood smear and no other info available. (PH26, note 2)

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## Primary Site Coding Instructions cont.

5. For extranodal lymphomas, secondary involvement [distant LNs, bone marrow, multifocal lung involvement, liver, spleen or CNS] are included in the staging fields only (not used for the purpose of coding primary site)
  - **EXCEPTION:** rare primary lymphoid neoplasms of spleen, liver or CNS (see PH Rules).
6. Items A-I (heme manual pages 33-38) provide instructions for coding the primary site for particular histologies

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## Items A-1: Primary Site by Histology

From Instruction 6 – Heme Manual pp. 36-39

Item	Code/PH Module	Primary Site	Histology
A	C379	Thymus	Thymic large B-cell lymphoma
	C383	Mediastinum	Mediastinal large B-cell lymphoma
B	C400-C419	Bone	Solitary plasmacytoma of bone
C	C420	Blood	Waldenstrom macroglobulinemia 2010-2017
D	C421	Bone Marrow	Long list, mostly leukemias
E	C422	Spleen	Splenic marginal zone lymphoma
			Hepatosplenic T-cell lymphoma
F	C44, C51, C60, C63	Skin sites	Skin lymphomas in list
G	C77 (unless extranodal)	Lymph Nodes	Mostly Hodgkin lymphomas
H	Module 3 and 4	PH5-PH8	Lymphoma/leukemias
I	Module 7		Lymphomas, extraosseous plasmacytomas, mast cell sarcomas, histiocytic and dendritic cell neoplasms, heavy chain disease, myeloid sarcoma, and PTLD

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## Histology Coding Instructions

1. Code histology ID'd by Definitive Diagnostic Method section of heme DB (no hierarchy)
  - Clinical diagnosis
  - Genetic test
  - Immunophenotyping
  - Cytology
  - Pathology
    - Final diagnosis
    - Comment on final diagnosis
    - Addenda to final diagnosis
    - CAP protocol/synoptic report
2. When Definitive Diagnostic Method tests or reports are not available, code from (in hierarchical order)
  - Documentation in the EMR
    - Referencing original scans, genetic testing, immunophenotyping, or pathology report
    - That refers to histology
  - Death certificate (central/regional registries only)

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## Histology Coding Instructions cont. (Instructions 3-5) Using Ambiguous Terms

3. Specific histology described with ambiguous term(s) plus "NOS" histology → code the NOS histology
  - Don't want to code a provisional dx that might change w/ further testing
  - If physician confirms the more specific histology, code more specific
- (4-5) ONE histology described by ambiguous term(s), Review Abstractor notes in Heme DB
  4. To see if other information can be used to confirm the dx
  5. For relevant immunophenotyping or genetics information that can be used to code that histology
    - Follow back to physician

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## PH Modules

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- Module 1: PTLD (PH1)
- Module 2: Plasmacytomas (9731/3, 9734/3) (PH2 – PH4)
- Module 3: CLL/SLL (9823/3) (PH5 – PH6)
- Module 4: Leukemia/Lymphoma (9727/3, 9811/3 – 9819/3, 9827/3, 9837/3) (PH7– PH8)
- Module 5: Myeloid Neoplasms and Mast Cell Neoplasms (multi histo codes) (PH9 – PH10)

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## PH Modules

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- Module 6: Non-Hodgkin Lymphomas (multi histo codes) (PH11 – PH17)
- Module 7: Hodgkin, NHL, and other histo (PH18 – PH27)
- Module 8: NOS & More Specific Histo (PH28 – PH29)
- Module 9: All heme and lymphoid neoplasms (9590/3 – 9992/3) (PH30 – PH31)

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## Appendices

Appendix A: *History of Hematopoietic and Lymphoid Neoplasm Coding*

Appendix B: *WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues*

Appendix C: *Lymph Node/Lymph Node Chain Reference Table*

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### Appendix C Lymph Node Chain Reference Table


Use with PH rules to determine whether involved nodes are in a single or multiple ICD-O LN regions (for the purpose of coding primary site)

- Alphabetic order
- ICD-O code
- ICD-O lymph node region
- AJCC lymph node region (for staging)

LN/LN Chain	ICD-O Code	ICD-O-3 LN Region	AJCC LN Region
Cloquet's node	C77.4	Inguinal region or leg	Inguino-femoral, right and left*
Colic NOS, ileocolic, mesocolic, middle (right)	C77.2	Intra-abdominal	Mesenteric
Common bile duct	C77.2	Intra-abdominal	Para-aortic
Cubital	C77.3	Axilla or arm	Axillary, right and left*
Cystic duct	C77.2	Intra-abdominal	Para-aortic

AJCC LN regions: see Figure 79.1, AJCC Cancer Staging Manual

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# Steps in Priority Order for Using the Heme DB and Hematopoietic Coding Manual

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## Using Heme DB and Coding Manual

1. Determine working histology by doing a SEARCH of the **Heme DB**
  - Leave the DB open while you complete the remaining steps
2. Determine number of primaries using the **M rules** - Start at M1
3. Verify or revise working diagnosis using the **PH rules**
4. Determine primary site using the **Heme DB** and **PH rules**
  - Some histologies can only have a single primary site; it will be listed in the Primary Site(s) field in the **Heme DB**
  - Review the Primary Site Text field in the **Heme DB**
    - Lists common primary sites of involvement (**Heme DB**)
    - Refers you to the appropriate PH module in the **PH rules**
    - Review Abstractor notes in **Heme DB**
    - Seek physician help
5. Code Grade for 2010-2017 cases (**Heme DB**)
  - When grade not provided in Heme DB, see **Heme manual**

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# Hematopoietic and Lymphoid Neoplasm Database

1 Search Database ICD-O-3 Code Lists 2 Downloads ▾

Show Multiple Primaries Calculator +

acute myelomonocytic 3 x Search

Show Alternate Names 4 (only comes up after you search)

69 neoplasms match any term 8 neoplasms match all terms 5 Show 25 Entries.

Relevance	ICD-O-3 Morphology	Name	Alternate Names
—	9867/3	<i>Acute myelomonocytic</i> leukemia	<i>Acute</i> myeloid leukemia, M4
—	9891/3	<i>Acute</i> monoblastic and monocytic leukemia	<i>Acute</i> monoblastic leukemia and <i>acute</i> monocytic leukemia <i>Acute</i> monoblastic leukemia <i>Acute</i> monoblastic leukemia, M5a <i>Acute</i> monocytic leukemia
—	9861/3	<i>Acute</i> myeloid leukemia, NOS	<i>Acute</i> granulocytic leukemia <i>Acute</i> myeloblastic leukemia <i>Acute</i> myelocytic leukemia <i>Acute</i> myelogenous leukemia

Search Database ICD-O-3 Code Lists

**Name**  
*Acute myelomonocytic* leukemia

**ICD-O-1 Morphology** Effective 1978 - 1991  
9867/3: Chronic granulocytic leukemia

**ICD-O-2 Morphology** Effective 1992 - 2000  
9867/3: *Acute myelomonocytic* leukemia

**ICD-O-3 Morphology** Effective 2001 and later  
9867/3: *Acute myelomonocytic* leukemia

**Reportable**  
for cases diagnosed 1978 and later

**Primary Site(s)**  
C421

**Help me code for diagnosis year :**  
2020

Coding Manual: Hematopoietic Coding Manual (PDF)

**Grade**

Not Applicable

**Module Rule**  
See abstractor notes

**Alternate Names**  
*Acute* myeloid leukemia, M4  
AMML  
AMMoL  
FAB M4

**Definition**  
*Acute myelomonocytic* leukemia is an *acute* leukaemic precursors. The peripheral blood or bone marrow has neutrophils and their precursors and monocytes in marrow cells.

**Abstractor Notes**  
If the leukemia occurs before or simultaneously with

**Definitive Diagnostic Methods**  
Bone marrow biopsy  
Immunophenotyping  
Peripheral blood smear

**Genetics Data**  
Gain of chromosome 8

**Immunophenotyping**

**Definition and Abstractor Notes** truncated to fit slide



# Staging

## AJCC 8<sup>th</sup> Edition

### EOD

### SS2018

### SSDs

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## AJCC Chapter, EOD Schema, and SS2018 Chapter

AJCC Chapter # and Name	EOD and SS2018	
71-Ocular Adnexal Lymphoma	Lymphoma Ocular Adnexa	
72-Brain and Spinal Cord (9680, 9714, 9702, 9712, 9699)	Brain	
	CNS Other	
79 - Hodgkin & NL Lymphoma	Lymphoma*	
80 - Pediatric Lymphoma	Lymphoma - CLL/SLL	
81-Primary Cutaneous Lymphoma	Primary Cutaneous Lymphoma: Non-MF/SS	
	Mycosis Fungoides and Sézary Syndrome	SS2018: Mycosis Fungoides
82-Plasma Cell Myeloma/Disorders	Plasma Cell Myeloma	SS2018: Myeloma and
	Plasma Cell Disorders	Plasma Cell Disorders
83-Leukemia	HemeRetic	

\*Excludes 9680, 9699, 9700-9714, 9751-9755 in CNS sites

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## AJCC 8<sup>th</sup> Summary of Changes

### Chapter 79 – Hodgkin & Non-Hodgkin Lymphoma

- Lugano classification (modification of Ann Arbor)
- B symptoms only used for Hodgkin lymphoma
- Eliminated
  - X subscript for bulk (record diameter of largest mass)
  - IIIIE (Now IV → extranodal involvement w/ nodal dz above & below diaphragm)
  - Stage IIIS: spleen involvement not part of staging
- Stage II: Bulky divided into limited or advanced, based on histology & prognostic factors
- P/A CXR no longer required for determining bulk (CT now used)

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## AJCC Classification System Summary

AJCC Chapter # and Name	AJCC Classification System
71-Ocular Adnexal Lymphoma	Uses T, N, & M; no Stage Group
72-Brain and Spinal Cord (9680, 9699, 9700-9714, 9751-9755)	No AJCC Staging
79-Hodgkin & NL Lymphoma	Hodgkin and NHL: Lugano
	CLL/SLL: Lugano and Rai
80-Pediatric Lymphoma	NHL: St. Jude
	Hodgkin: Lugano
81-Primary Cutaneous Lymphoma	Non-MF and SS: Uses T, N, & M; no Stage Group
	MF and SS: ISCL/EORTC (uses T, N, M, peripheral blood involvement, & Stage Group)
82-Plasma Cell Myeloma/Disorders	RISS Stage Group
83-Leukemia (CLL - In Lymphoma Chapter)	AML - Disseminated at dx (no anatomic staging)
	ALL - Disseminated at dx (no anatomic staging)
	ALL (pediatric) - Based on age and WBC count
	CML - Based on BM morphology & cytogenetic changes

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## Hodgkin and Non-Hodgkin Lymphoma Staging SS2018 and EOD

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- Summary Stage (only 4 applicable codes)
  - 1 Localized only
  - 2 Regional, NOS
  - 7 Distant site(s)/lymph nodes involved
  - 9 Unknown if extension or metastasis
- EOD Primary Tumor
  - codes 100, 300, and 600 are only used for **nodal** lymphomas
  - code 200 is reserved for **extranodal** lymphomas (designated in *italics* on the next slide)
  - Codes 400, 600, 700, 750, and 800 can be used for **either** nodal or extranodal lymphomas
- EOD Regional Nodes – only code is 888
- EOD Mets – only code is 88

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## Mets at Diagnosis Fields 2021+

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- Mets at Dx Bone, Brain, Liver, Lung, Distant LNs, Other
  - Assign for these histologies included in the HemeRetic schema
    - Dendritic neoplasms (9756/3-9759/3)
    - Erdheim-Chester Disease (9749/3-new histology for 2021)
    - Langerhans cell histiocytosis, disseminated (9751/3)
    - Mast cell sarcoma (9740/3)
    - Myeloid Sarcoma (9930/3)
  - When EOD Primary Tumor is
    - 100, there are no mets at dx (code all to 0)
    - 700-800 and SS is code 7, there could be mets
- Use code 8 (Not applicable) for the following
  - Any case coded to primary site C420, C421, C423, C424. (C770-C779  
Mets at Dx - Distant LNs; Code the remaining Mets at Dx fields)
  - Plasma Cell Disorders 00822

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## Hodgkin and Non-Hodgkin Lymphoma Staging SS2018 and EOD, cont

- Descriptions are divided into nodal and extranodal lymphomas (improves readability of the tables)
- EOD code (750) for peripheral blood involvement ONLY
- Descriptions updated and clarified
  - Example]
    - (v1.7) Diffuse or disseminated (multifocal) involvement of ONE OR MORE extralymphatic organ(s)/site(s)
    - (v2.0) Diffuse or disseminated involvement (except multifocal lung involvement or any liver involvement, see code 800) of MORE than one extralymphatic organ/site

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## Hodgkin and Non-Hodgkin Lymphoma Staging

N/EN	EOD	SS18	Description
N	100	1	Single lymph node region involved
			Involvement of multiple nodal chains in the SAME lymph node region
EN	200		Single extralymphatic site <b>W/O</b> nodal involvement
			Multifocal involvement (except lung or any liver involvement) of <b>1</b> extralymphatic organ/site <b>W/O</b> nodal involvement
N	300		Two or more lymph node regions involved <b>SAME</b> side of diaphragm
N	400		Contiguous extralymphatic extension from nodal/lymphatic site <b>W/</b> or <b>W/O</b> involvement of other nodal regions on <b>SAME</b> side of diaphragm (i.e., LN extending to extralymphatic site)
EN			Localized involvement of a single extralymphatic organ/site <b>W/</b> involvement of its RLN(s) OR <b>W/</b> involvement of other LN(s) on <b>SAME</b> side of the diaphragm
N	500		Bulky disease present (EOD: W/ codes 300 or 400; AJCC: II + bulk)
N/EN	600	7	Involvement of LN regions on BOTH sides of the diaphragm OR nodes ABOVE the diaphragm involved W/ spleen involvement

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## Hodgkin & Non-Hodgkin Lymphoma Staging

EOD	SS18	Description	
700	7	Diffuse or disseminated involvement (except multifocal lung or any liver involvement) of 1 or > 1 extralymphatic organ(s)/site(s) <b>W/</b> or <b>W/O</b> associated LN involvement	
		Involvement of isolated extralymphatic organ in absence of involvement of adjacent LN(s), but in conjunction w/ dz in distant sites	
		Multifocal involvement (except multifocal lung or any liver involvement) of 1 extralymphatic organ/site <b>W/</b> nodal involvement	
		Noncontiguous extralymphatic organ involvement in conjunction <b>W/</b> nodal disease (2 or more sites involved)	
750		Peripheral blood involvement ONLY	
800		Diffuse or disseminated involvement of bone or CNS	
		Bone marrow	Any Involvement
		Cerebrospinal fluid (CSF)	
		Liver	
		<b>Multiple</b> lung lesions (other than by direct extension in SS18 code 2, EOD code 400)	
Peripheral blood involvement w/ other involvement			

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## Hodgkin and Non-Hodgkin Lymphoma AJCC Staging

- AJCC 8<sup>th</sup> Edition separates into limited and advanced stage based on:
  - Number of sites involved
  - Location of involved sites in relation to diaphragm
  - Spleen involvement
  - Liver, bone marrow, CSF, multiple lung lesions

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## SSDI: B Symptoms

- MD Statement can be used when no other info
- Pruritis alone, ETOH intolerance, fatigue, or fever associated w/ a suspected infection do not qualify for B classification
- AJCC only uses B symptoms for classification of Hodgkin Lymphoma

Code	Description
0	No B symptoms (asymptomatic) Classified as "A" by physician when asymptomatic
1	Any B symptom(s) Night sweats (drenching – enough to require change of bedclothes) Unexplained fever (above 38 degrees C) Unexplained weight loss (>10% of body wt in the 6 months before dx) B symptoms, NOS Classified as "B" by physician when symptomatic
8	Not applicable: Information not collected for this case
9	Not documented in medical record B symptoms not assessed or unknown if assessed

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## SSDI: HIV Status

- MD Statement can be used when no other info
- AIDS-associated lymphomas are a late manifestation of HIV infection & have unique clinical & pathological features that differ from lymphomas in the general population (extranodal involvement w/ CNS most common site)
- If patient has a history of HIV, assign code 1 even if HIV is not currently detectable.

Code	Description
0	Not associated with Human Immunodeficiency Virus (HIV)/Acquired Immune Deficiency Syndrome (AIDS) HIV negative
1	Associated with HIV/AIDS HIV positive
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 8 will result in an edit error.)
9	Not documented in medical record HIV status not assessed or unknown if assessed

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## SSDI: NCCN International Prognostic Index

- MD statement of NCCN IPI (points or risk) **must** be used
  - Applicable for non-Hodgkin lymphomas only (use X9 for HL)
- A low, intermediate or high risk associated with a RAI Stage is not recorded in this data item.

Code	Description
00-08	0-8 points (points have priority over risk when both are available)
X1	Stated as low risk (0-1 point)
X2	Stated as low intermediate risk (2-3 points)
X3	Stated as intermediate risk (4-5 points)
X4	Stated as high risk (6-8 points)
X8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code X8 will result in an edit error.)
X9	Not documented in medical record NCCN International Prognostic Index (IPI) not assessed or unknown if assessed

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## Pediatric Hodgkin and Non-Hodgkin Lymphoma AJCC Chapter 80

- Hodgkin lymphoma uses Lugano classification (staged the same way as adult Hodgkin lymphoma)
- Non-Hodgkin lymphoma uses the St. Jude Children's Research Hospital system (stages I, II, III, and IV and based on number of sites involved and location/extent of involvement)

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## CLL/SLL 9823/3 AJCC Staging

- Staged as a lymphoma
  - Lugano **and** Rai staging have to be assigned
    - Lugano in AJCC fields
    - Rai in SSDI
- Primary site
  - PH5, if bone marrow or peripheral blood involved, code site to C42.1
    - AJCC stage IV due to + bone marrow
  - PH6, if NO bone marrow (or unknown) or NO peripheral blood involvement, code site to LN, LN region, organ, or tissue of origin
    - AJCC stage depends on description of tissues involved

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## CLL/SLL 9823/3 Rai Staging

### SSDIs used for Rai Stage

#### Adenopathy

LN > 1.5cm?

#### Organomegaly

Liver/spleen large?

#### Anemia

Hgb < 11.0 g/dL?

#### Lymphocytosis

Lymphocytes > 5,000 cells/ $\mu$ L?

#### Thrombocytopenia

Platelets < 100,000  $\mu$ L?

### Modified Rai Staging System

RAI Stage	Findings	Survival (mo)
0	Lymphocytosis only	> 120
I	+ Adenopathy	95
II	+ Enlarged spleen and/or liver	72
III	Lymphocytosis + Hgb < 11 g/dL	30
IV	Lymphocytosis + Platelets < 100,000/ $\mu$ L	30

#### SSDI Code Structure

All 5 have 0, 1, 5, & 9

Lab values also have 6 & 7

0 – absent

1 – present

5 – Not applicable; Primary site not C421

6 – present per MD, lab value unknown

7 – ordered, results not in chart

9 – Not documented; unknown if assessed

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## Lab Value SSDIs: CLL/SLL 9823/3 Rai Staging

Code	[Lymphocytosis]	[Anemia]	[Thrombocytopenia]
0	Not present		
	ALC <= 5,000 cells/μL	Hgb >=11.0 g/dL Physician states Rai stage 0-II	Platelets (Plt) >=100,000/μL Physician states Rai stage 0-III
1	Present		
	ALC > 5,000 cells/μL	Hgb <11.0 g/dL	Platelets (Plt) < 100,000/μL
5	Not applicable: Primary site is not C421		
6	Lab value unknown, physician states [ ] is present		
	Physician states Rai stage 0-IV	Physician states Rai stage III	Physician states Rai stage IV
7	Test ordered, results not in chart		
9	Not documented in medical record		
	[ ] not assessed or unknown if assessed		
	No Rai stage is documented in the record and there is no documentation of [ ]		
		Physician states Rai stage IV and there is no documentation of anemia	

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## Adenopathy and Organomegaly SSDIs: CLL/SLL 9823 Rai Staging

Code	[Adenopathy]	[Organomegaly (Hepato- or spleno-megaly)]
0	[ ] not identified/not present	
	No lymph nodes > 1.5 cm	
	Physician states Rai stage 0	Physician states Rai stage 0-I
1	Present	
	Presence of lymph nodes > 1.5 cm	
	Physician states Rai stage I	Physician states Rai stage II
5	Not applicable: Primary site is not C421	
9	Not documented in medical record	
	[ ] not assessed or unknown if assessed	
	No Rai stage is documented in the record and there is no documentation of [ ]	
	Physician states Rai stage II-IV and there is no documentation of adenopathy	Physician states Rai stage III-IV and there is no documentation of organomegaly

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## HEME/RETIC

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### CASE 1

- A 61-year old male, previously healthy.
- April 2019, knee injury, followed by deep vein thrombosis of the left leg and pulmonary embolism. He completed 6 months of Eliquis.
- **Past medical history:** none
- **Past surgical history:** none
- **Family history:** unremarkable
- **Travel history:** no foreign travel in past 3 months
- November 2020, his CBC showed worsening cytopenia.

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## CASE 1, continued.

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### Workup

- 12/14/2020 absolute neutrophil count 0.73, hemoglobin 9.3, platelet count 63, absolute total white cell count 0.8; in 2 weeks his white cell count dropped from 2.4 to 0.82.
- 12/3/2020: AML: Blasts 40% of nucleated cells; CD45+, CD34-, CD 117+, CD13 positive, CD33 positive in 59.6%, HLA-DR dim, and myeloperoxidase dim; Cytogenetics normal karyotype; **Next generation sequencing detected IDH 2 p.(R172K)c515>A.**

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## CASE 1, continued.

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### Treatment for AML

- Cycle #1 Vidaza for 7 days from 12/17/21- 12/24/21 and Venetoclax 100 mg daily from 12/24/2021 onwards
  - The dose of Venetoclax is reduced because he is taking Posaconazole 300 mg daily for fungal prophylaxis
- Cycle #2 Vidaza for 7 days- 1/18/2021- 1/26/21. Venetoclax stopped on 2/16/21
- 1/2021 Restaging marrow after cycle #1 Azacitadine + Venetoclax: Patient is in complete remission. No evidence of IDH 1, IDH 2 mutation
- 3/2021 Marrow post Aza + Venetoclax #2 shows complete remission, IDH2 NOT detected
- 03/2021: Allogeneic Stem cell transplant for AML

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aml Search

Show Alternate Names

35 neoplasms match Show 25 Entries.

▲ Relevance	ICD-O-3 Morphology	Name	Alternate Names
—	9861/3	Acute myeloid leukemia, NOS	Acute myeloid leukemia with cytoplasmic nucleophosmin (NPMc + AML) AML
—	9920/3	Therapy-related myeloid neoplasms	t-AML t-AML/t-MDS
—	9898/3	Myeloid leukemia associated with Down Syndrome	AML associated with DS
—	9912/3	Acute myeloid leukemia with BCR-ABL1	AML with BCR-ABL1
—	9878/3	Acute myeloid leukemia with biallelic mutation of CEBPA	
—	9895/3	Acute myeloid leukemia with myelodysplasia-related changes	AML-MRC
—	9874/3	Acute myeloid leukemia with maturation	AML with maturation

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## Heme Database Abstractor Notes for AML, NOS

### Abstractor Notes

- Acute myeloid [leukemia](#), NOS is a [generic](#) disease description. [DCO cases](#) or path report only [cases](#) may stay in this classification.

In most [cases](#), NOS [histology](#) is only the provisional [diagnosis](#); the physician will run further [diagnostic procedures](#) and look for various clinical presentations to identify a more specific disease. Further review of the [medical record](#) should be done to look for the tests listed as [definitive diagnosis](#). If no information is found in the [medical record](#), [follow-back](#) to the attending physician should be done.

- Physician clarification:** “The WHO category for this AML was: AML with recurrent genetic abnormalities.”

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# Hematopoietic and Lymphoid Neoplasm Coding Manual

- **Rule M2** Abstract a single primary when there is a single histology.
  - We have a single histology, AML with recurrent genetic abnormalities
  
- **Note 3:** A single histology is diagnosed by the definitive diagnostic method as defined in the Heme DB. For example, the patient had several provisional diagnoses but the definitive diagnostic method identified a single histology. Abstract as a single primary.


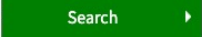
95


recurrent ← x Search

Show Alternate Names ← Pre-2022 updates!

8 neoplasms match Show 25 ▾ Entr









▲ Relevance	ICD-O-3 Morphology	Name	Alternate Names
—	9718/1	Lymphomatoid papulosis	
—	9895/3	Acute myeloid leukemia with myelodysplasia-related changes	
—	9896/3	Acute myeloid leukemia, t(8;21)(q22;q22.1); RUNX1-RUNX1T1	Acute myeloid leukemia with <span style="background-color: yellow;">recurrent</span> genetic abnormalities
—	9725/3	Hydroa vacciniforme-like lymphoma <span style="background-color: #cccccc;">obsolete</span>	
—	9871/3	Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFβ-MYH11	
—	9762/3	Heavy chain diseases	
—	9877/3	Acute myeloid leukemia with mutated NPM1	
—	9725/1	Hydroa vacciniforme like lymphoproliferative disorder	96

recurrent  

Show Alternate Names 


**Post-2022 updates!**

8 neoplasms match Show  Entries.

▲ Relevance	ICD-O-3 Morphology	Name	Alternate Names
	9861/3	Acute myeloid leukemia, NOS	Acute myeloid leukemia with <b>recurrent</b> genetic abnormalities, NOS
	9718/1	Lymphomatoid papulosis	
	9895/3	Acute myeloid leukemia with myelodysplasia-related changes	
	9725/3	Hydroa vacciniforme-like lymphoma <small>obsolete</small>	
	9871/3	Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFβ-MYH11	
	9762/3	Heavy chain diseases	
	9877/3	Acute myeloid leukemia with mutated NPM1	
	9725/1	Hydroa vacciniforme like lymphoproliferative disorder	


- Primary site: C42.1
- Histology: 9861/3

[Search Database](#) [ICD-O-3 Code Lists](#)

**Name**  
Acute myeloid leukemia, NOS 


**ICD-O-1 Morphology** Effective 1978 - 1991  
9861/3: Acute myeloid leukemia, NOS

**ICD-O-2 Morphology** Effective 1992 - 2000  
9861/3: Acute myeloid leukemia, NOS

**ICD-O-3 Morphology** Effective 2001 and later  
9861/3: Acute myeloid leukemia, NOS 

**Reportable**  
for cases diagnosed 1978 and later

**Primary Site(s)**  
C421  
Primary site must be bone marrow (C421)

 **Help me code for diagnosis year :**  
2020

## Grade

Not Applicable

## Module Rule

See abstractor notes

## Alternate Names

Acute [granulocytic leukemia](#)

Acute [myeloblastic leukemia](#)

Acute [myelocytic leukemia](#)

Acute [myelogenous leukemia](#)

Acute myeloid [leukemia](#), NOS

[Acute myeloid leukemia](#), NOS (FAB or WHO type not specified)

[Acute myeloid leukemia](#) with [cytoplasmic nucleophosmin \(NPMc + AML\)](#)

[Acute myeloid leukemia](#) with [mutated CEBPA](#)

[Acute myeloid leukemia](#) with [mutated NPM1](#)

[Acute myeloid leukemia](#) with recurrent [genetic abnormalities](#), NOS

[Acute non-lymphocytic leukemia](#)

[AML](#)

FAB M0

99

## Definitive Diagnostic Methods Immunophenotyping

Bone marrow biopsy

CD11b

Genetic testing

CD13

Immunophenotyping

CD14

## Genetics Data

CD33 MPO

CEBPA

CD34

FLT3-ITD

CD68

NPM1

Varies by subclassification

12/3/2020: AML: Blasts 40% of nucleated cells. CD45 positive, CD34 negative, CD 117+, CD13 positive, CD33 positive in 59.6% and HLA-DR was dim and myeloperoxidase was dim. Cytogenetics normal karyotype. **The next generation sequencing detected IDH 2 p.(R172K)c515>A.**

100

# Diagnostic Confirmation

Microscopically Confirmed – Use Codes 1-4

Code	Description
1	Positive histology • Includes: peripheral blood smear only
2	Positive cytology
3	Positive histology <b>PLUS</b> : • Positive immunophenotyping AND/OR • Positive genetic studies • Includes: peripheral blood smear followed by flow cytometry <i>(Effective for cases diagnosed 1/1/2010 and later)</i>
4	Positive microscopic confirmation, method not specified

101

Schema: HemeRetic

## Grade Table 88

Schema ID#	Schema ID Name	AJCC ID	AJCC Chapter
00790	Lymphoma	79.0-79.4, 79.6	Hodgkin and Non-Hodgkin Lymphoma
00795	Lymphoma-CLL/SLL	79.5	Hodgkin and Non-Hodgkin Lymphoma
00811	Mycosis Fungoides	81.1	Primary Cutaneous Lymphoma: Mycosis Fungoides and Sezary Syndrome
00812	Primary Cutaneous Lymphomas (excluding Mycosis Fungoides)	81.2	Primary Cutaneous Lymphoma: B-Cell/T-cell Lymphoma (non-MF/SS) Lymphoma
00821	Plasma Cell Myeloma	82.1	Plasma Cell Myeloma and Plasma Cell Disorders
00822	Plasma Cell Disorders	82.2	Plasma Cell Myeloma and Plasma Cell Disorders
00830	HemeRetic	83.0-83.4	Leukemia

**Note:** Grade (cell indicator) is no longer applicable for this hematopoietic neoplasm.

Code	Grade Description
8	Not applicable

102



**Grade Clinical**

(Cannot be left blank)

8

**Grade Pathological**

(Cannot be left blank)

8

**Grade Post Therapy Clinical**

Leave blank when no neoadjuvant therapy)

Blank

**Grade Post Therapy Pathological**

(Leave blank when no neoadjuvant therapy)

Blank

**EOD Primary Tumor**

**Notes**

- Note 1 lists histologies that can be localized (code 100), systemic (700) or unknown (999)
- Note 2: LN involvement for histologies included in Note 1 is not collected
- Note 3 lists histologies are systemic (code 700)

**We assign code 700 per the list of histologies in Note 3 that are always systemic.**

Code	Description
100	Localized disease
	(Single/solitary/unifocal/isolated)
	See Notes 1 and 2
700	Systemic disease
	See Note 3
999	Unknown; extension not stated
	Primary tumor cannot be assessed
	Not documented in patient record
	Death Certificate Only

## Schema: HemeRetic

### EOD Regional Nodes

Code	Description
888	Not applicable: Information not collected for this schema

### EOD Mets

Code	Description
88	Not applicable: Information not collected for this schema

105

## Schema: HemeRetic

### Summary Stage 2018

#### Notes

- Note 3 lists histologies that can be localized (code 1), systemic (7) or unknown (9)
- Note 4 states LN involvement for histologies included in Note 1 is not collected
- Note 5 lists histologies are systemic (code 7)

We assign code 7 per the list of histologies in Note 3 that are always systemic.

Code	Description
1	Localized disease
	(Single/solitary/unifocal/isolated)
	See Notes 2 and 3
7	Distant site(s)/Lymph nodes(s) involved
	Systemic disease
	See Note 4
9	Unknown if extension or metastasis

106

**Schema: HemeRetic****SSDI: JAK2**

- **Note 1:** Physician statement of JAK2 can be used to code this data item when no other information is available.
- **Note 2:** Janus Kinase 2 (JAK2, JAK 2) is a gene mutation that increases susceptibility to several myeloproliferative neoplasms (MPNs). Testing for the JAK2 mutation is done on whole blood. Nearly all people with polycythemia vera, and about half of those with primary myelofibrosis and essential thrombocythemia, have the mutation.
- **Note 3:** Record JAK2 for any hematopoietic neoplasm. It is most commonly used for the following histologies:
  - Polycythemia Vera (9950/3)
  - Primary myelofibrosis (9961/3)
  - Essential Thrombocytopenia (9962/3)
  - Chronic myelomonocytic leukemia (9945/3)

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**Schema: HemeRetic****SSDI: JAK2**

Code	Description
0	JAK2 result stated as negative
1	JAK2 positive for mutation V617F WITH or WITHOUT other mutations
2	JAK2 positive for exon 12 mutation
3	JAK2 positive for other specified mutation
4	JAK2 positive for more than one mutation other than V617F
5	JAK2 positive NOS Specific mutation(s) not stated
7	Test ordered, results not in chart
8	Not applicable: Information not collected for this case (If this item is required by your standard setter, use of code 8 will result in an edit error.)
9	Not documented in medical record JAK2 not assessed or unknown if assessed

108



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## PLASMA CELL MYELOMA

109



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### CASE 2

- 76 yr old male
- Past medical history: type 2 diabetes, hypertension, prostatomegaly
- Past surgical history: hernia repair
- Family history: mother had CLL
- Travel history: no foreign travel in past 3 years
- September 2012:
  - Presented with lethargy and fatigue to PCP
  - Initial CBC, showed Hb of 9.4gm/dl, WCC of 4.7 and platelets-137,000
  - Chemistry results showed normal renal function, but increased total protein with increased globulin of 7.2gm/dl

110

## CASE 2, continued.

---

Referred to Hem/Onc

### Workup

- SPEP showed monoclonal protein
- Multiple lytic bone lesions

### September 2012: Stage II IgG lambda myeloma

- Staging: M spike 3.0; Beta 2 macroglobulin 4.87; Hgb 14.1; Ca 16.1; Cr 1.32; Albumin 4.4; BM Inadequate sample; 4% flow; Normal cytogenetics

111

## CASE 2, continued

---

### Treatment for Myeloma

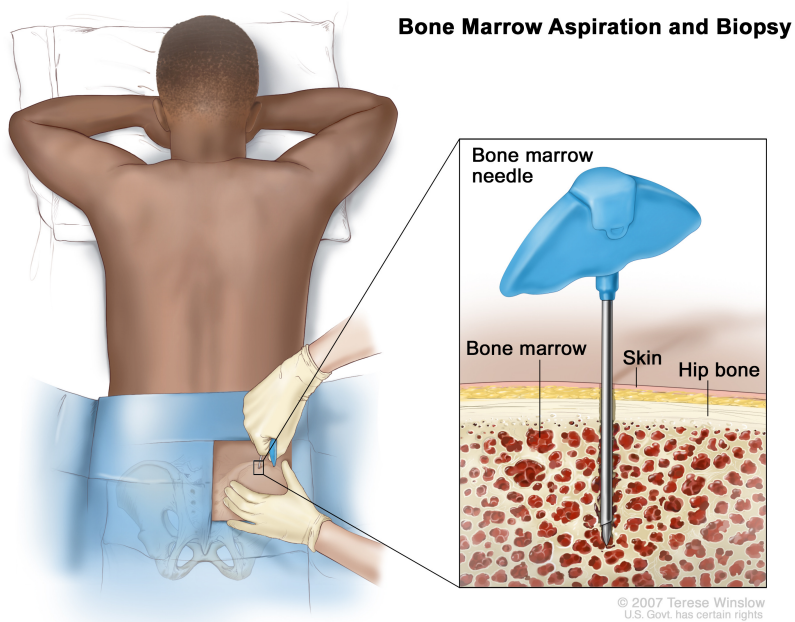
- **First Line Therapy:** RVD (VD X1;RVD X3) VGPR; Cyt/G-CSF mobilization
- **Melphalan auto PBSCT 3/6/2013; Revlimid maintenance until Aug 2017**
- **June 2017:** BM 35% plasma cells; M spike 1.7; lambda FLC 135
- **August-November 2017:** Elotuzomab/Revlimid/Dexamethasone for 3 cycles
- **November 2017** Increasing lambda FLC progression

112

## CASE 2, continued.

- **November 2017-May 2018:** Daratumumab/pomalidomide/Dex (24 weeks); Re-Staging: June 19, 2018; Pre-HCT evaluation; Very Good Partial Remission documented
- **July 2018** Auto #2 with Mel 140
- **09/2018-05/20** Pomalidomide maintenance (stopped with increasing PSA)
- Weekly Velcade maintenance
- **10/26/20:** Progression of Myeloma Lambda was 758.8 with M-spike 0.8
- **11/02/20** Lambda 791.7, M-spike 1.4
- **11/12/20:** Started Darzalex/Velcade dex cycle 1 day 1

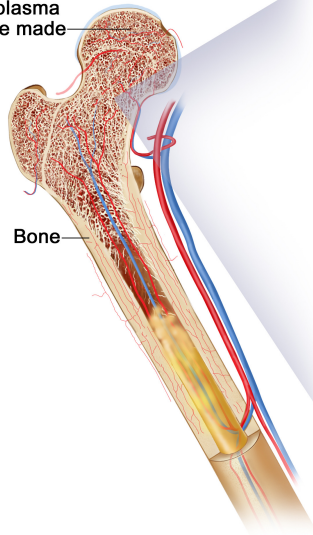
113



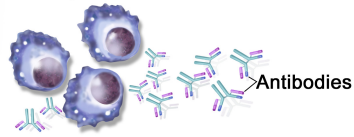
114

## Multiple Myeloma

Red marrow where plasma cells are made

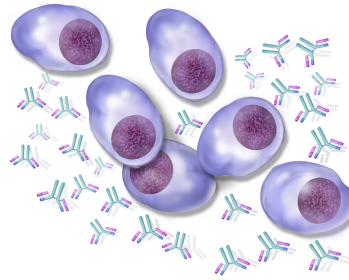


Normal plasma cells



Antibodies

Multiple myeloma cells (abnormal plasma cells)



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115

myeloma ✕ Search ▶

Show Alternate Names

11 neoplasms match Show 25 Entries.

▲ Relevance	ICD-O-3 Morphology	Name	Alternate Names
—	9732/3	Plasma cell <b>myeloma</b>	Alpha plasma cell <b>myeloma</b> Early multiple <b>myeloma</b> Early <b>myeloma</b> Evolving multiple <b>myeloma</b>
—	None	Plasma cell neoplasm	
—	9731/3	Solitary plasmacytoma of bone	Solitary <b>myeloma</b>
—	None	Evolving myelodysplasia	
—	9734/3	Extrasosseous plasmacytoma	
—	9733/3	Plasma cell leukemia <small>obsolete</small>	
—	9765/1	Non-IgM monoclonal gammopathy of undetermined significance	Non- <b>myelomatous</b> gammopathy

116

**Primary Site:**

**C42.1  
Bone Marrow**

**Histology:**

**9732/3  
Plasma Cell  
Myeloma**

Search Database ICD-O-3 Code Lists

**Name**  
Plasma cell *myeloma* ←

**ICD-O-1 Morphology** Effective 1978 - 1991  
9730/3: Plasma cell *myeloma*  
9830/3: Plasma cell leukemia

**ICD-O-2 Morphology** Effective 1992 - 2000  
9732/3: Plasma cell *myeloma*

**ICD-O-3 Morphology** Effective 2001 and later  
9732/3: Plasma cell *myeloma* ←

**Reportable**  
for cases diagnosed 1978 and later

**Primary Site(s)**  
C421  
Primary site must be bone marrow (C421) ←

**Help me code for diagnosis year :**  
2012 ←

117

**Hematopoietic and Lymphoid Neoplasm Coding Manual**

- **Rule M2** Abstract a single primary when there is a single histology.
  - We have a single histology, plasma cell myeloma

Example 1: The diagnosis is multiple myeloma (9732/3). Abstract as a single primary.

118



## Definitive Diagnostic Methods

### Definitive Diagnostic Methods

Bence-Jones protein  
 Bone marrow biopsy  
 FISH  
 Genetic testing  
 Immunophenotyping  
 Peripheral blood smear  
 Serum Protein Electrophoresis (SPEP)

### Immunophenotyping

CD19-  
 CD38  
 CD56 aberrantly expressed (except PCL)  
 CD56- (PCL)  
 CD79a  
 CD138  
 VS38c

### Genetics Data

Five major **oncogenes** involved in 14q32 **translocation**: cyclin D1, C-MAF, FGFR3/MMSET, cyclin D3, and MAFB  
 High load of IGHV **gene somatic** hypermutation  
 Immunoglobulin heavy and **light chain genes** are clonally rearranged  
 Trisomies  
 Whole or partial **chromosome deletions** or **translocations**

119

## Diagnostic Confirmation

Not Microscopically Confirmed – Use Codes 5-8

Code	Description
5	Positive laboratory test/marker study Note 1: Includes cases with positive immunophenotyping or genetic studies and no histological confirmation Note 2: This does not include cases where a peripheral blood smear is done (code 1) and peripheral blood smear followed by flow cytometry (code 3)
6	Direct visualization without microscopic confirmation
7	Radiology and other imaging techniques without microscopic confirmation
8	Clinical diagnosis only (other than 5, 6 or 7)

**We assign code 5 based on the SPEP.**

120

## Grade (Pre 2018)

6 - B-cell

## Module Rule

None

## Alternate Names

Alpha PCM

Alpha plasma cell myeloma

Early multiple myeloma

Early myeloma

Evolving multiple myeloma

Evolving myeloma

Evolving plasma cell myeloma

Gamma PCM

Gamma plasma cell myeloma

Indolent myeloma

Indolent PCM

Indolent plasma cell myeloma

Kahler's disease

Medullary plasmacytoma

From this point, we will pretend this is a 2021 diagnosis so that we can show you the fields that are currently being abstracted.

Multiple myeloma

Multiple plasmacytomas (occurring in bone or outside of bone)

Myeloma, NOS

Myelomatosis

Non-secretory myeloma

PCL

PCM

Plasma cell leukemia

Plasmacytic leukemia

Primary PCL

Secondary plasma cell leukemia

Smoldering myeloma

Smoldering plasma cell myeloma

121

## Schema: Plasma Cell Myeloma

## Grade Table 88

Schema ID#	Schema ID Name	AJCC ID	AJCC Chapter
00790	Lymphoma	79.0-79.4, 79.6	Hodgkin and Non-Hodgkin Lymphoma
00795	Lymphoma-CLL/SLL	79.5	Hodgkin and Non-Hodgkin Lymphoma
00811	Mycosis Fungoides	81.1	Primary Cutaneous Lymphoma: Mycosis Fungoides and Sezary Syndrome
00812	Primary Cutaneous Lymphomas (excluding Mycosis Fungoides)	81.2	Primary Cutaneous Lymphoma: B-Cell/T-cell Lymphoma (non-MF/SS) Lymphoma
00821	Plasma Cell Myeloma	82.1	Plasma Cell Myeloma and Plasma Cell Disorders
00822	Plasma Cell Disorders	82.2	Plasma Cell Myeloma and Plasma Cell Disorders
00830	HemeRetic	83.0-83.4	Leukemia

**Note:** Grade (cell indicator) is no longer applicable for this hematopoietic neoplasm.

Code	Grade Description
8	Not applicable

122

**Schema: Plasma Cell Myeloma**

**Grade Fields**

**Grade Clinical**

(Cannot be left blank)

8

**Grade Pathological**

(Cannot be left blank)

8

**Grade Post Therapy Clinical**

Leave blank when no neoadjuvant therapy)

Blank

**Grade Post Therapy Pathological**

(Leave blank when no neoadjuvant therapy)

Blank

**Abstractor Notes**

Plasma cell **myeloma** (PCM) usually has generalized **bone marrow** involvement. **Lytic bone lesions** and **bone tumor masses** of **plasma cells** also occur.

Approximately 30% of patients with solitary **plasmacytoma** (**bone** or outside of **bone**) defined only by radiographical skeletal **survey** have additional **lesions** identified on **MRI** or **CT**. These patients are considered to have **plasma cell myeloma**.

The International **Staging System** for **Multiple Myeloma** **Staging** for **Multiple Myeloma** is based on:

1. Amount of monoclonal (or **myeloma**) **protein** (**M protein**) in the **serum** and/or **urine**
2. Various clinical **parameters** such as: **hemoglobin** and **serum calcium** concentrations, number of **lytic bone lesions**
3. Presence or absence of **renal failure**.

Stage I

Stage II

Stage III

**Schema: Plasma Cell Myeloma**

**EOD Primary Tumor**

**Note:** 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 700** for systemic involvement.

Code	Description
700	Multiple myeloma (9732) Myeloma, NOS (9732) Plasma cell myeloma (9732)
999	Unknown; extension not stated Primary tumor cannot be assessed Not documented in patient record Death Certificate Only

**We assign code 700 based on the diagnosis of myeloma.**

125

**Schema: Plasma Cell Myeloma**

**EOD Regional Nodes**

Code	Description
888	Not applicable: Information not collected for this schema

**EOD Mets**

Code	Description
88	Not applicable: Information not collected for this schema

126

**Schema: Plasma  
Cell Myeloma**

**Summary Stage**

**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.

SS2018	Description
1	Localized only <ul style="list-style-type: none"> <li>• Single plasmacytoma occurring in bone (osseous or medullary) (9731)W/ or W/OUT soft tissue extn.</li> <li>• Single plasmacytoma, NOS (9734)                             <ul style="list-style-type: none"> <li>• Single plasmacytoma occurring outside of bone (extraosseous or extramedullary) (9731)</li> </ul> </li> </ul>
3	Regional lymph nodes only <ul style="list-style-type: none"> <li>• Extraosseous plasmacytomas only (9734)                             <ul style="list-style-type: none"> <li>• Regional lymph node(s), NOS                                     <ul style="list-style-type: none"> <li>• Lymph node(s), NOS</li> </ul> </li> </ul> </li> </ul>

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**Schema: Plasma  
Cell Myeloma**

**Summary Stage**

SS2018	Description
7	Distant site(s)/lymph node(s) involved <ul style="list-style-type: none"> <li>•Lymphoplasmacytic lymphoma (9671)</li> <li>•Waldenstrom Macroglobulinemia (9761)</li> <li>•Plasma cell myeloma (9732)                             <ul style="list-style-type: none"> <li>• Multiple myeloma</li> <li>• Myeloma, NOS</li> <li>• Multiple extraosseous or extramedullary plasmacytomas</li> <li>• Multiple osseous or medullary plasmacytomas</li> <li>• Multiple plasmacytomas, NOS</li> </ul> </li> </ul>
9	Unknown if extension or metastasis (applicable for 9731 and 9734 only)

**We assign code 7 per Note 3.**

128

## Plasma Cell Myeloma - AJCC Staging

- AJCC 8<sup>th</sup> Edition bases staging on
    - Serum beta macroglobulin
    - Serum albumin
    - High risk cytogenetics
    - LDH
  - Collected as SSDI
  - No AJCC staging for myeloma in 2012
  - If 2018+ diagnosis, AJCC stage II
- Beta 2 macroglobulin 4.87  
Albumin 4.4  
Cytogenetics Normal  
No LDH

129

## Plasma Cell Myeloma - SSDI Schema Discriminator 1: Multiple Myeloma Terminology

Instructions for Coding	Code	Description	Staging
<p><b>Note 1:</b> Terms in HEME DB effective 1/1/2010 and later</p> <p><b>Note 2:</b> Select the terminology specified by the physician in the record, NOT based on criteria in AJCC 8<sup>th</sup> table 82.1</p> <p><b>Note 3:</b> Do not change this code if term used later indicates progression</p> <p><b>Note 4:</b> If plasma cell leukemia dx WITH plasma cell myeloma, <b>code 5</b></p>	0	Multiple myeloma Myeloma, NOS Non-secretory myeloma Plasma cell myeloma (PCM) Ultra-high-risk smoldering MM (SMM)	RISS Stage
	1	Asymptomatic plasma cell myeloma Early myeloma Evolving myeloma Smoldering plasma cell myeloma (SPCM)	NO RISS Stage
	9	Other terminology describing myeloma Unknown terminology used	NO RISS Stage

**We assign code 0 based on the physician statement of myeloma.**

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## Plasma Cell Myeloma - SSDI High Risk Cytogenetics

**Note 1:** Physician statement of presence/absence can be used to code this item

**Note 2:** Code physician statement or FISH test interpretation at dx (pre-tx)

**Note 3:** If test results differ from physician statement, physician statement takes precedence

**Note 4:** If no mention of high-risk cytogenetics, code 9

**Note 5:** If schema discriminator 1 coded to 1 or 9, **assign code 5**

Code	Description
0	High-risk cytogenetics not identified/not present
1	High-risk cytogenetics present
5	Schema discriminator 1: Plasma Cell Myeloma Terminology coded to 1 or 9
7	Test ordered, results not in chart
9	Not doc. in medical record; high risk cytogenetics not/unk if assessed

**High Risk Cytogenetics is defined as one or more: t(4;14), t(14;16), or del 17p**

**We assign code 0 based on the physician statement of normal cytogenetics.**

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## Plasma Cell Myeloma SSDI Serum Albumin Pretreatment Level

**Note 1:** Elevated serum albumin is  $\geq 3.5$  g/dL. (If lab is grams per liter (g/L) that is 10 times of g/dL.  $3.5$  g/dL =  $35$  g/L)

**Note 2:** Based on **blood** test pre-tx; physician statement of exact value can be used in the absence of the lab test; do NOT use findings from urine test

**Note 3:** If no mention of serum albumin, code 9

**Note 4:** If schema discriminator 1 coded to 1 or 9, **assign code 5**

Code	Description
0	Serum albumin $< 3.5$ g/dL
1	Serum albumin $\geq 3.5$ g/dL
5	Schema discriminator 1: Plasma Cell Myeloma Terminology coded to 1 or 9
7	Test ordered, results not in chart
9	Not doc. in medical record; serum albumin pretx level not assessed or unk if assessed

**Albumin is the most abundant protein in human plasma.**

**We assign code 1 based on the physician statement of 4.4.**

132



## Plasma Cell Myeloma SSDI Serum Beta-2 Microglobulin Pretreatment Level

Note 1: Part of RISS staging; use cut points in table regardless of lab's reference range  
 Note 2: Based on **blood** test pre-treatment; physician statement of exact value can be used in the absence of the lab test; use the highest value available  
 Note 3: If no mention of  $\beta$ 2-microglobulin, code 9  
 Note 4: If schema discriminator 1 coded to 1 or 9, **assign code 5**

Code	Description
0	$\beta$ 2-microglobulin < 3.5 mg/L
1	$\beta$ 2-microglobulin $\geq$ 3.5 mg/L < 5.5 mg/L
2	$\beta$ 2-microglobulin $\geq$ 5.5 mg/L
5	Schema discriminator 1: Plasma Cell Myeloma Terminology coded to 1 or 9
7	Test ordered, results not in chart
9	Not doc. in medical record; serum albumin pretx level not assessed or unk if assessed

**Serum  $\beta$ 2-microglobulin is protein on surface of many cells and plentiful on WBC surface; increased production or destruction of cells causes  $\beta$ 2-microglobulin level to increase**

**We assign code 1 based on the physician statement of 4.87.**

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## Plasma Cell Myeloma SSDI LDH Pretx Level (Lactate Dehydrogenase)

Note 1: Use the reference ranges from your lab to determine if LDH is normal  
 Note 2: Based on **blood** test pre-tx; physician statement of exact value can be used in the absence of the lab test; use the highest value available  
 Note 3: If no mention of LDH, code 9  
 Note 4: If schema discriminator 1 coded to 1 or 9, **assign code 5**

Code	Description
0	Normal LDH level; low, below normal
1	Above normal LDH level; high
5	Schema discriminator 1: Plasma Cell Myeloma Terminology coded to 1 or 9
7	Test ordered, results not in chart
9	Not documented in medical record; LDH pretx level not/unk if assessed

**LDH is an enzyme involved in conversion of sugars to energy and present in most cells in the body.**

**We assign code 9 since there is no mention of LDH.**

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## SSDI for Heme

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### HemeRetic

- Jak2

### Plasma Cell Myelomas

- High Risk Cytogenetics
- Serum Alb Pre-Tx Level
- Serum Beta-2 Microglobulin Pre-Tx Level
- LDH Pre-Tx Level
- Mycosis Fungoides
  - Peripheral Blood Involvement

### Lymphoma

- B Symptoms
- HIV Status
- NCCN IPI

### CLL/SLL

- B Symptoms
- HIV Status
- NCCN IPI
- Adenopathy
- Organomegaly
- Anemia
- Lymphocytosis
- Thrombocytopenia

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## Questions about...

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- Heme manual and DB
  - **SINQ**
    - Public Q&A
    - Updated yearly?
    - Searchable
  - **Ask a SEER Registrar**
    - Private email answers
    - Possibility it could be added to SINQ
  - **This presentation**
    - [DeniseCHarrisonLLC@gmail.com](mailto:DeniseCHarrisonLLC@gmail.com)
    - [LouanneCurrence@NKCH.org](mailto:LouanneCurrence@NKCH.org)
- **Staging and SSDI**
  - **CAnswer Forum**
    - Public Q&A
    - Updated?
    - Searchable

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**Thank you!**

It's time to take some questions.

## Fabulous Prizes



5

## Coming UP...

- Colon 2022
  - Guest: Janice Smith, CTR
  - 5/05/2022
- Central Nervous System 2022
  - Jim Hofferkamp, CTR
  - 6/02/2022

6

## CE Certificate Quiz/Survey

CE Phrase

Link



7

**Thank you!**

- [jhofferkamp@naaccr.org](mailto:jhofferkamp@naaccr.org)
- [amartin@naaccr.org](mailto:amartin@naaccr.org)



8

8