### **LEUKEMIA**

Includes Acute Lymphocytic Leukemia (ALL), Chronic Lymphocytic Leukemia (CLL), Acute Myeloid Leukemia (AML) and Chronic Myeloid Leukemia (CML)

### **Background**

This case definition was developed by the Armed Forces Health Surveillance Branch (AFHSB) for the purpose of descriptive epidemiological reports on invasive cancers among active duty Service members.<sup>1</sup>

### **Clinical Description**

Leukemia is a cancer of the bone marrow and blood and is classified into four main groups according to cell type and rate of growth: acute lymphocytic (ALL), chronic lymphocytic (CLL), acute myeloid (AML), and chronic myeloid (CML). Almost 90% of cases are diagnosed in adults 20 years of age or older, with the most common types being AML and CML. In children, ALL is the most common type. Symptoms include lymphadenopathy, night sweats, and recurrent infections. The cause of leukemia is unknown; however, risk factors for developing the condition include exposure to ionizing radiation, benzene exposure (particularly with AML), and heredity for CLL. Treatment varies depending on the type and stage of disease. Leukemia was estimated to be the cause of about 3% of new cancer cases and 4% of all cancer deaths in the United States in 2012.<sup>2</sup>

#### **Case Definition and Incidence Rules**

For surveillance purposes, a case of leukemia is defined as:

- One hospitalization with a case defining diagnosis of leukemia (see ICD9 and ICD10 code lists below) in the first diagnostic position; or
- One hospitalization with a V or Z-code indicating a radiotherapy, chemotherapy, or immunotherapy treatment procedure (see ICD9 and ICD10 code lists below) in the *first* diagnostic position; AND any case defining diagnosis of leukemia (see ICD9 and ICD10 code lists below) in the *second* diagnostic position; or
- Three or more outpatient medical encounters, occurring within a 90-day period, with a case defining diagnosis of leukemia (see ICD9 and ICD10 code lists below) in the *first* or second diagnostic position.

#### Incidence rules:

For individuals who meet the case definition:

(continued on next page)

<sup>&</sup>lt;sup>2</sup> American Cancer Society. Cancer Facts & Figures 2019. Atlanta: American Cancer Society; 2019.



<sup>&</sup>lt;sup>1</sup> Armed Forces Health Surveillance Center. Incident diagnoses of cancers and cancer-related deaths, active component, U.S. Armed Forces, 2005-2014. *Medical Surveillance Monthly Report (MSMR)*. 2016 July; 23(7): 23-31.

# Case Definition and Incidence Rules (continued)

• The incidence date is considered the date of the first hospitalization or the first of the three or more outpatient medical encounters occurring within a 90-day period that includes a case defining diagnosis of leukemia (see *Case Definition and Incidence Rule Rationale*).

• An individual is considered an incident case only *once per lifetime*.

# **Exclusions:**

None

# Codes

The following ICD9 and ICD10 codes are included in the case definition:

Condition	ICD-10-CM Codes	ICD-9-CM Codes
Lymphoid leukemia	C91 (lymphoblastic leukemia)	204 (lymphoid leukemia)
	C91.0 (acute lymphoblastic leukemia)	204.0 (lymphoid leukemia, acute)
	- C91.00 (not having achieved remission)	- 204.00 (w/o mention of having achieved remission or failed remission)
	- C91.01 (in remission)	- 204.01 (in remission)
	- C91.02 (in relapse)	- 204.02 (in relapse)
	C91.1 (chronic lymphocytic leukemia of B-cell type)	204.1 (lymphoid leukemia, chronic)
	- C91.10 (not having achieved remission)	- 204.10 (w/o mention of having achieved remission or failed remission)
	- C91.11 (in remission)	- 204.11 (in remission)
	- C91.12 (in relapse)	- 204.12 (in relapse)
	C91.3 (prolymphocytic leukemia of B-cell type)	204.8 (other lymphoid leukemia)
	- C91.30 (not having achieved remission)	- 204.80 (w/o mention of having achieved remission or failed remission)
	- C91.31 (in remission)	- 204.81 (in remission)
	- C91.32 (in relapse)	- 204.82 (in relapse)
	C91.4 (hairy cell leukemia)	- 202.40 (leukemic reticuloendotheliosis,
	- C91.40 (not having achieved remission)	unspecified site, extranodal and solid organ sites)
	- C91.41 (in remission)	

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	- C91.42 (in relapse)	
	C91.5 ( <i>adult T- cell</i> lymphoma/leukemia (HTLV-1-associated)	- 204.8[0-2] (above)
	- C91.50 (not having achieved remission)	
	- C91.51 (in remission)	
	- C91.52 (in relapse)	
	C91.6 ( <i>prolymphocytic</i> leukemia of T-cell type)	- 204.8[0-2] (above)
	- C91.60 (not having achieved remission)	
	- C91.61 (in remission)	
	- C91.62 (in relapse)	
	C91.A ( <i>mature B-cell</i> leukemia Burkitt-type)	- 204.8[0-2] (above)
	- C91.A0 (not having achieved remission)	
	- C91.A1 (in remission)	
	- C91.A2 (in relapse)	
	C91.Z (other lymphoid leukemia)	204.2 (lymphoid leukemia, subacute)
	- C91.Z0 (not having achieved remission)	- 204.20 (w/o mention of having achieved remission or failed remission)
	- C91.Z1 (in remission)	- 204.21 (in remission)
	- C91.Z2 (in relapse)	- 204.22 (in relapse)
	C91.9 (lymphoid leukemia, <i>unspecified</i> )	204.9 (unspecified lymphoid leukemia)
	- C91.90 (not having achieved remission)	204.90 (w/o mention of having achieved remission or failed remission)
	- C91.91 (in remission)	- 204.91 (in remission)
	- C91.92 (in relapse)	- 204.92 (in relapse)
Myeloid	C92 (myeloid leukemia)	205 (myeloid leukemia)
leukemia	C92.0 (acute myeloblastic leukemia)	205.0 (myeloid leukemia, acute)
	- C92.00 (not having achieved remission)	- 205.00 (w/o mention of having achieved remission or failed remission)
	- C92.01 (in remission)	- 205.01 (in remission)
	- C92.02 (in relapse)	- 205.02 (in relapse)
	C92.1 (chronic myeloid leukemia, BCR/ABL-positive)	205.1 (myeloid leukemia, chronic)
	- C92.10 (not having achieved remission)	- 205.10 (w/o mention of having achieved remission or failed remission)

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- C92.11 (in remission)	- 205.11 (in remission)
- C92.12 (in relapse)	- 205.12 (in relapse)
C92.2 (atypical chronic myeloid leukemia, BCR/ABL-negative)	205.2 (myeloid leukemia, subacute)
- C92.20 (not having achieved remission)	- 205.20 (w/o mention of having achieved remission or failed remission)
- C92.21 (in remission)	- 205.21 (in remission)
- C92.22 (in relapse)	- 205.22 (in relapse)
C92.3 (myeloid sarcoma)	205.3 (myeloid sarcoma)
- C92.30 (not having achieved remission)	- 205.30 (w/o mention of having achieved remission or failed remission)
- C92.31 (in remission)	- 205.31 (in remission)
- C92.32 (in relapse)	- 205.32 (in relapse)
C92.4 (acute promyelocytic leukemia)	205.0[0-2] (above)
- C92.40 (not having achieved remission)	
- C92.41 (in remission)	
- C92.42 (in relapse)	
C92.5 (acute myelomonocytic leukemia)	205.0[0-2] (above)
- C92.50 (not having achieved remission)	
- C92.51 (in remission)	
- C92.52 (in relapse)	
C92.6 (acute myeloid leukemia with 11q23 abnormality)	205.0[0-2] (above)
- C92.60 (not having achieved remission)	
- C92.61 (in remission)	
- C92.62 (in relapse)	
C92.A (acute myeloid leukemia with multilineage dysplasia)	205.0[0-2] (above)
- C92.A0 (not having achieved remission)	
- C92.A1 (in remission)	
- C92.A2 (in relapse)	
C92.Z (other myeloid leukemia)	205.8 (other myeloid leukemia)
- C92.Z0 (not having achieved remission)	- 205.80 (w/o mention of having achieved remission or failed remission)
- C92.Z1 (in remission)	- 205.81 (in remission)

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	- C92.Z2 (in relapse)	- 205.82 (in relapse)
	C92.9 (myeloid leukemia, unspecified)	205.9 (unspecified myeloid leukemia)
	C92.90 (not having achieved remission)	- 205.90 (w/o mention of having achieved remission or failed remission)
	C92.91 (in remission)	- 205.91 (in remission)
	C92.92 (in relapse)	- 205.92 (in relapse)
Monocytic	C93 (monocytic leukemia)	206 (monocytic leukemia)
leukemia	C93.0 (acute monoblastic/monocytic leukemia)	206.0 (monocytic leukemia, acute)
	- C93.00 (not having achieved remission)	- 206.00 (w/o mention of having achieved remission or failed remission)
	- C93.01 (in remission)	- 206.01 (in remission)
	- C93.02 (in relapse)	- 206.02 (in relapse)
	C93.1 (chronic myelomonocytic leukemia)	206.1 (monocytic leukemia, chronic)
	- C93.10 (not having achieved remission)	- 206.10 (w/o mention of having achieved remission or failed remission)
	- C93.11 (in remission)	- 206.11 (in remission)
	- C93.12 (in relapse)	- 206.12 (in relapse)
	C93.3 ( <i>juvenile</i> myelomonocytic leukemia)	206.8 (other monocytic leukemia)
	- C93.30 (not having achieved remission)	- 206.80 (w/o mention of having achieved remission or failed remission)
	- C93.31 (in remission)	- 206.81 (in remission)
	- C93.32 (in relapse)	- 206.82 (in relapse)
	C93.Z (other monocytic leukemia)	206.8[0-2] (above)
	- C93.Z0 (not having achieved remission)	
	- C93.Z1 (in remission)	
	- C93.Z2 (in relapse)	
	C93.9 (monocytic leukemia, <i>unspecified</i> )	206.2 (monocytic leukemia, <i>subacute</i> )
		206.9 (unspecified monocytic leukemia)
	- C93.90 (not having achieved remission)	- 206.20 (w/o mention of having achieved remission or failed remission)
		- 206.90 (w/o mention of having achieved remission or failed remission)
	- C93.91 (in remission)	- 206.21 (in remission) - 206.91 (in remission)
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	- C93.92 (in relapse)	- 206.22 (in relapse)
	(accompany)	- 206.92 (in relapse)
Other specified	C94 (other specified leukemia)	207 (other specified leukemia)
leukemia	C94.0 (acute erythroid leukemia)	207.0 (other specified leukemia, <i>acute</i> erythremia and erythroleukemia)
	- C94.00 (not having achieved remission)	- 207.00 (w/o mention of having achieved remission or failed remission)
	- C94.01 (in remission)	- 207.01 (in remission)
	- C94.02 (in relapse)	- 207.02 (in relapse)
	C94.2 (acute megakaryoblastic leukemia)	207.2 (megakaryocytic leukemia)
	- C94.20 (not having achieved remission)	- 207.20 (w/o mention of having achieved remission or failed remission)
	- C94.21 (in remission)	- 207.21 (in remission)
	- C94.22 (in relapse)	- 207.22 (in relapse)
	C94.3 (mast cell leukemia)	207.8 (other specified leukemia)
	- C94.30 (not having achieved remission)	- 207.80 (w/o mention of having achieved remission or failed remission)
	- C94.31 (in remission)	- 207.81 (in remission)
	- C94.32 (in relapse)	- 207.82 (in relapse)
	C94.8 (other specified leukemias)	207.8[0-2] (above)
	- C94.80 (not having achieved remission)	
	- C94.81 (in remission)	
	- C94.82 (in relapse)	
Leukemia of	C95 (leukemia of unspecified cell type)	208 (leukemia of unspecified cell type)
unspecified cell type	C95.0 ( <i>acute</i> leukemia of unspecified cell type)	208.0 (leukemia of unspecified cell type, acute)
	- C95.00 (not having achieved remission)	- 208.00 (w/o mention of having achieved remission or failed remission)
	- C95.01 (in remission)	- 208.01 (in remission)
	- C95.02 (in relapse)	- 208.02 (in relapse)
	C95.1 (chronic leukemia of unspecified cell type)	208.1 (leukemia of unspecified cell type, chronic)
	- C95.10 (not having achieved remission)	- 208.10 (w/o mention of having achieved remission or failed remission)
	- C95.11 (in remission)	- 208.11 (in remission)
	- C95.12 (in relapse)	- 208.12 (in relapse)

C95.9 (leukemia, unspecified)	208.2 (leukemia of <i>unspecified cell type</i> , subacute)
	208.8 ( <i>other</i> leukemia of unspecified cell type)
	208.9 (unspecified leukemia)
- C95.90 (not having achieved remission)	- 208.[2,8,9]0 (w/o mention of having achieved remission or failed remission)
- C95.91 (in remission)	- 208.[2,8,9]1 (in remission)
- C95.92 (in relapse)	- 208.[2,8,9]2 (in relapse)

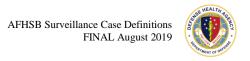
Procedures	ICD-10-CM Codes	ICD-9-CM Codes
Related treatment procedures	Z51.0 (encounter for antineoplastic radiation therapy)	V58.0 (radiotherapy)
(Radiotherapy, chemotherapy, immunotherapy)	Z51.1 (encounter for antineoplastic chemotherapy and immunotherapy)	V58.1 (encounter for chemotherapy and immunotherapy for neoplastic conditions)
	- Z51.11 (encounter for antineoplastic chemotherapy)	V58.11 (encounter for antineoplastic chemotherapy)
	- Z51.12 (encounter for antineoplastic immunotherapy)	V58.12 (encounter for antineoplastic immunotherapy)

### **Development and Revisions**

- In April of 2017 the case definition was updated to include ICD10 codes.
- This case definition was developed in 2010 by the Armed Forces Health Surveillance Center (AFHSC) in collaboration with a working group of subject matter experts from the Office of the Assistant Secretary of Defense for Health Affairs (ASDHA), the United States Army Public Health Command (USAPHC) and the United States Military Cancer Institute. The case definition was developed based on reviews of the ICD9 codes, the scientific literature, and previous AFHSC analyses.
- This case definition was developed for a report on ten different invasive cancers. As such, the same case finding criteria are used for all types of cancer in the report. This broad application of a single case definition may affect the sensitivity and specificity in varying ways for the individual cancers. Furthermore, surgical treatment procedures such as hysterectomy, mastectomy, prostatectomy, and other procedures unique to certain types of cancer are not included in the code sets for individual cancers.

# Case Definition and Incidence Rule Rationale

- Case finding criteria for this definition requires one hospitalization record with a case-defining ICD9 or ICD10 code for leukemia in the *first* diagnostic position *unless* a code for a related treatment procedure is in the *first* diagnostic position; then the case-defining ICD9 or ICD10 code for leukemia is allowed in the *second* diagnostic position.
- The case finding criterion of three or more outpatient medical encounters, within a 90-day period, with a case defining diagnosis of leukemia is used to identify cases that do not meet the other criteria



in the definition. For outpatient encounters, the incident date is considered the first of the three encounters occurring within the 90-day period (e.g., if an individual has four leukemia codes on 1 Jan 12, 1 Dec 15, 8 Dec 15, and 15 Dec 15, the incident date would be 1 Dec 15. 1 Jan 12 would be considered a screening encounter and dropped). Exploratory analysis of the Defense Medical Surveillance System (DMSS) data revealed that this criterion yielded optimal specificity. The period of 90 days was established to allow for the likelihood that "true" cases of leukemia would have second and third encounters within that interval.

For the purposes of counting new incident cases, AFHSB uses a once per lifetime incidence rule unless
a specific timeframe is more appropriate and is specified (e.g., individuals may be counted as an
incident case once every 365 days). Historically, a "once per surveillance period" incidence rule was
used due to limited data in the Defense Medical Surveillance System (DMSS), but that is no longer
necessary.

### Code Set Determination and Rationale

• In April of 2017 ICD9 codes 207.1x (chronic erythemia) / ICD10 D45 (polycythemia vera) were removed from the code set. The disorders are conditions related to the overproduction of red blood cells and, in general, not related to leukemia.

#### Reports

AFHSB reports on leukemia in the following reports:

• Periodic *MSMR* articles on cancers and cancer-related deaths.

Review	
Aug 2019	Case definition reviewed and updated by the AFHSB Surveillance Methods and Standards (SMS) working group.
Apr 2017	Case definition reviewed and updated by the AFHSB Surveillance Methods and Standards (SMS) working group.
Apr 2013	Case definition reviewed and adopted by the AFHSC Surveillance Methods and Standards (SMS) working group.
Jun 2012	Case definition developed and adopted by the AFHSC MSMR staff.
Comments	

None

<sup>&</sup>lt;sup>3</sup> Detailed information on this analysis is available through AFHSB *MSMR* staff; reference DMSS Requests #R080127, #R080159, #R090184, #R090302, #R090341, #R100181, and #R100303 (DoD Cancer Incidence), 2008-2009.