



NSW Oncology Group Haematological Cancer Minimum Dataset Extension

Data Dictionary

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Introduction

The purpose of this document is to define the key indicators and data elements for haematology, to complement the demographic, treatment and quality of care elements captured through clinical cancer registration. This data set proposal has arisen from discussion in the Minimum Data Set subcommittee of the NSW Greater Metropolitan Clinical Taskforce (GMcT) Bone Marrow Transplant (BMT) Clinicians Group, and was developed by Dr Campbell Tiley, haematologist at the Northern Sydney & Central Coast Area Health Service.

This dataset adds the additional elements needed to classify and monitor treatment outcomes for patients with haematological malignancies.

For each haematological malignancy, the ICD 10AM¹ diagnosis codes and morphology codes are used for classification. The World Health Organisation morphology codes have been incorporated into the ICD10AM taxonomy (the “M codes”), and those pertinent to haematological malignancies are listed in Appendix II.

Table 1: Summary of Haematological Data Items

1	Cancer diagnosis	The ICD-10-AM code representing this specific disease. This is an existing data item in cancer registry collections. In these collections the data item is called site of primary cancer. This item will be used to determine the haematological cancer group to which this cancer belongs. The haematological cancer group is not a new data item, it is a subgroup – or specialization determined from the data held in the cancer diagnosis field.
The following items may be recorded for a given cancer diagnosis. The elements to be used are determined by the type of cancer, therefore not every cancer type requires each of these items. When ever entered, they have an associated date.		
2	Staging	Staging or disease status classification data elements are specific to the staging system suggested for the type of cancer. The data element therefore indicates the system used and the value held in that item indicates the score obtained at a given reporting date.
3	Prognostic Score	A new data element recording the prognostic score for this patient’s haematological cancer is entered and related to a date.
4	Response	A new data element to record outcomes of therapy (response and treatment failure) with multiple entries possible per patient each of which is related to a date.
5	Prognostic Marker	This is a potential data item that can record prognostically important markers. Usually one per patient disease but more than one may be recorded. Marker detail (list of options) Marker Value: Pos, Neg, Not Done

Addition of data items (3, 4 above) would substantially enhance the utility of the Clinical Cancer Registry for documenting clinically relevant outcomes in Haematological malignancies. Item 5 would substantially enhance the ability of the registry to incorporate a number of highly sensitive molecular and cytogenetic prognostic factors. The number of such markers is likely to expand in the immediate future and can be incorporated in this data structure without use of additional data elements.

It is anticipated that

- These items are added to the NSW Minimum Data Set for clinical cancer registration, where a patient has a haematological malignancy
- That ongoing management of the haematology dataset will be the responsibility of the NSW Haematology Group convened by the Cancer Institute NSW.

¹ International Classification of Disease, Australian Modification

Background

Cancer Notification

Cancer is a notifiable disease under the NSW Public Health Act (1991)². The NSW Central Cancer Registry holds cancer notifications from 1972, and is operated by the Cancer Institute NSW.

The purpose of the Central Cancer Registry is to register all *incident* cancer cases, and all cancer *deaths*. The primary output is the annual *Cancer in NSW: Incidence and Mortality report*³.

In addition, the Registry provides a source of enrolment in clinical trials and other cancer studies, subject to strict privacy considerations and Ethics Committee approval. Five-year survival rates, which are the major measurements of treatment effectiveness when controlled for disease stage, are generally sourced from the NSW Central Cancer Registry.

The primary sources of notifications are NSW hospitals, including radiotherapy facilities, and laboratories. The NSW Registry of Births Deaths and Marriages provides death notifications to the Registry, and the Australian Bureau of Statistics provides *coded* death certificates.

Currently (2003 figures), the Registry receives around 170,000 notifications, which translate to approximately 31,400 cancer cases in the state. There are around 12,000 cancer deaths annually in NSW.

Table 2: Cancer Notification

Facility	Referring doctor, hospital/laboratory
Person	Identification, age, sex, country of birth, place of residence
Disease	Identified by the clinician and recorded in the existing registry using the code for primary site of cancer and cancer morphology fields.

Clinical Cancer Registration

To enhance the epidemiological information provided by the NSW Central Cancer Registry, the Cancer Institute NSW is working with Area Health Services to pilot a clinical minimum data set for cancer, based on NSW⁴ and national standards⁵. This dataset, also referred to as *clinical registration*, adds the dimension of *stage*, *treatment* and *quality of care* to what is known about incidence and mortality⁶.

² For all NSW legislation, see <http://www.legislation.nsw.gov.au/maintop/scanact/inforce/NONE/0>

³ See Tracey EA, Roder D, Bishop J, Chen S, Chen W, Cancer in New South Wales: Incidence and Mortality 2003. Sydney: Cancer Institute NSW, 2005. The latest report (1993) is available at: http://www.cancerinstitute.org.au/cancer_inst/statistics/pdfs/IncidenceMortalityReport2005.pdf.

⁴ see http://www.cancerinstitute.org.au/cancer_inst/statistics/registryccr.html

⁵ Health Data Standards Committee 2004. Data Set Specification, Cancer (clinical), National Health Data Dictionary Version 12, Supplement. AIHW Cat. No. HWI 71. Canberra: Australian Institute of Health and Welfare

⁶ More detail on the NSW clinical cancer registry pilot is available at http://www.cancerinstitute.org.au/cancer_inst/statistics/registryccr.html

Table 3: Clinical Cancer Registration

Stage	Tumour, nodes, metastases (TNM) OR clinical staging system, basis for staging. This includes staging systems specific to haematological malignancies
Treatment	Surgery, anti-neoplastic agents, radiation
Quality of Care	Psychosocial assessment, clinical trials enrolment, multidisciplinary team involvement

Disease-Specific Information

Patterns of care studies collect a broader and finer level of information on patients treated within given disease groups. Typically, they assess the compliance of a cohort of cancer patient treatments against best practice guidelines.

However, it may be possible to leverage the increasingly sophisticated hospital administrative, clinical and laboratory systems to complement clinical cancer registration with a succinct, critical set of indicators for one disease group regularly, rather than sporadically.

The next generation NSW cancer registry will address:

- Current statutory reporting requirements, including a migration to message-and-extract-based notification.
- Clinical cancer registration – the collection of a minimum data set for all cancer cases in NSW, describing stage and treatment.
- Disease-specific indicators and surveys, which will be collected intermittently or regularly, for specialty cancer groups. For instance, gynaecology oncology, genitourinary, upper gastrointestinal, haematopoietic malignancies.

The registry will continue to collect information on site and morphology of disease and these existing data items will be used to define the haematological cancer group to which the patient belongs. There will be no need to collect additional information in order to define the cancer group.

Some NSW Oncology Groups are already identifying a succinct set of indicators specific to each disease group. Before developing a data collection strategy, this phase of the Institute's NSW clinical cancer information strategy involves development and documentation of data standards to support each disease-specific stream. This set of indicators pertains to the following haematological malignancies (haematological cancer group):

- Hodgkin Lymphoma (HD)
- Non-Hodgkin Lymphoma (NHL) – with the following specific sub-sets
 - Follicular Lymphoma
 - Aggressive Non-Hodgkin Lymphoma
 - Other Non-Hodgkin Lymphoma
- Acute Myeloid Leukaemia (AML)
- Acute Lymphoblastic Leukaemia (ALL)
- Chronic Myeloid Leukaemia (CML)
- Chronic Lymphocytic Leukaemia (CLL)
- Multiple Myeloma (MM)
- Myelodysplastic Syndromes (MDS)

For these diseases, the following classifications are incorporated into the data dictionary for the NSW Minimum Data Set for clinical cancer registration. Any of these items may have a date associated with them. This date can be compared to the diagnosis date for the specific cancer. Such an approach allows multiple entry and the recording of variations over time for all data items.

Table 4: Haematological Domains of Existing MDS items

Cancer diagnosis (derived from existing data)	Use of ICD10-AM to identify the haematological cancer group to which this entry belongs. This item therefore uses the existing data item of cancer diagnosis to identify the cancer group. This is an item derived from the existing field for cancer diagnosis, both site and morphology and need not be an additional data entry.
Staging	This field includes stage of disease or classification of progress. Alternative staging classifications currently incorporated in the dictionary are: Ann Arbor for Hodgkin's and Non-Hodgkin Lymphoma Chronic Myeloid Leukaemia Stage Durie and Salmon Stage for Myeloma Binet Stage for Chronic Lymphocytic Leukaemia Rai Stage for Chronic Lymphocytic Leukaemia

The NSW Haematology Group proposes that the following items are adopted as the standard additional items for haematological cancer registration.

Table 5: Key Haematological Measures

Prognostic Score	Prognostic score is a value to indicate the prognostic score according to a defined scoring approach. Defined values include: Follicular Lymphoma International Prognostic Index (FLIPI) International Prognostic Index (IPI) Hasenclever Score for Hodgkin Lymphoma International Staging System for Multiple Myeloma. International Prognostic Scoring System for Myelodysplastic Syndrome Sokal Score for Chronic Myeloid Leukaemia Cytogenetic Prognostic Group for Acute Myeloid Leukaemia In some cases classifications that are called staging systems have been included in this area as they represent prognostic information, despite their name.
Response	This item indicates responses to treatment, including failures. The data structure allows this information to be recorded with an associated date to allow investigation of response over time.

Addition of the data items in Table 5 would substantially enhance the utility of the Clinical Cancer Registry for documenting clinically relevant outcomes in haematological malignancies.

A further two items would substantially enhance the ability of the registry to incorporate a number of highly sensitive molecular and cytogenetic prognostic factors. The number of such markers is likely to expand in the immediate future and can be incorporated in this data structure without use of additional data elements.

Table 6: Additional Prognostic Markers

Prognostic Marker	Propose incorporation of prognostically important markers, predominantly cytogenetic and molecular. Usually one per patient, possibly multiple. Marker type (list of options) Marker Value: Pos, Neg, Not Done (possibly incorporate technique here eg: FISH, qPCR etc) Date Recorded
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Layout of this Document

This document includes detailed data definitions and background information organized in the following manner:

Data Dictionary General Concepts

Specific Haematological malignancies and common data items for all groups.

Value Domains including staging systems, prognosis and responses for each group.

- Hodgkin Lymphoma (HD)

- Non-Hodgkin Lymphoma (NHL) – with the following specific sub-sets

 - Follicular Lymphoma

 - Aggressive Non-Hodgkin Lymphoma

 - Other Non-Hodgkin Lymphoma

- Acute Myeloid Leukaemia (AML)

- Acute Lymphoblastic Leukaemia (ALL)

- Chronic Myeloid Leukaemia (CML)

- Chronic Lymphocytic Leukaemia (CLL)

- Multiple Myeloma (MM)

- Myelodysplastic Syndromes (MDS)

Structure

The data dictionary includes two different types of things. The data elements into which information is recorded. For example: In the case of the data element cancer diagnosis the value domain is ICD10-AM. There may be more than one data element that uses a specific value domain, for example there are many data elements that use ICD10-AM as their value domain, including principle diagnosis and cancer diagnosis. ICD10-AM is often referred to as WHO classification. Once a value domain is described it can be referenced by as many data elements as are appropriate. In this document there are several value domains that are used by more than one data element, for example 'Stage of Disease (Ann Arbor)'. It is for this reason that the value domains are listed specifically in Appendix 2, the value domain appendix, and described in less detail in the body of the document.

In the past data dictionaries simply listed the data elements. This generated a great deal of repetition and lack of consistency. The current national approach to data element specification acknowledges that items exist in a structure that increases their ability to accurately represent the information intended. The structure suggested for haematological cancer data is shown in Figure 1.

Each element that is lower in the tree 'belongs' to the item under which they are shown. This structure means that each individual cancer diagnosis may have multiple instances of cytogenetic and molecular markers (as this group of data items are linked directly to the cancer diagnosis field group). Each cancer diagnosis can be identified as belonging to one of the specific haematological cancer groups (all shown below the cancer diagnosis field group) see Table 1. Each of these groups can be determined using values from existing elements of the data collection. These data values are indicated in table 14.

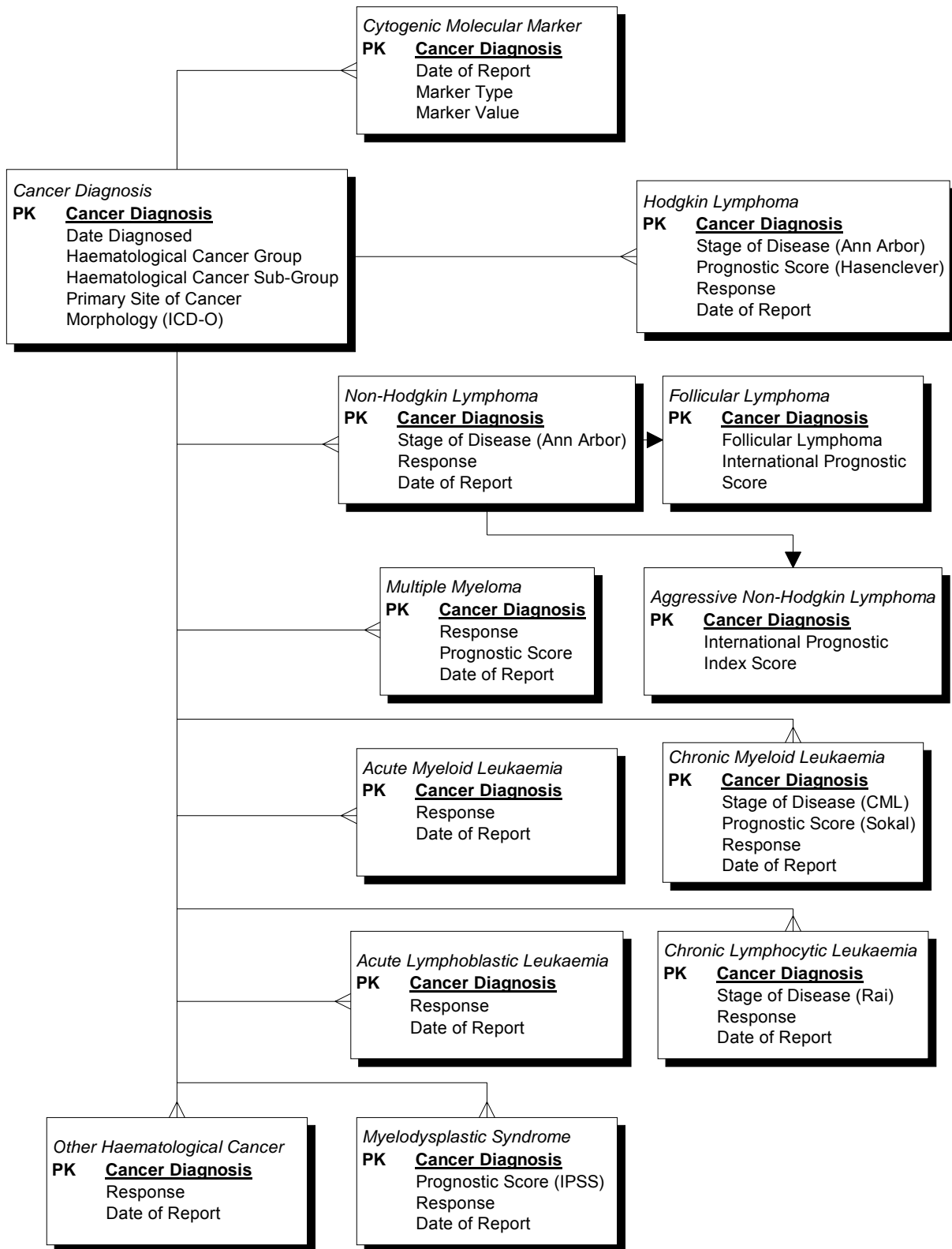
The Other Haematological Cancer group indicates the data elements used by haematological cancers that do not have specific requirements for recording of staging or prognosis. These items will still be diagnostically identifiable, and include response and date information and markers where appropriate but do not require more specific data elements.

For example: In Figure 1 the data entity for Non-Hodgkin Lymphoma has two sub entities – (1) for Follicular Lymphoma and (2) for Diffuse Non-Hodgkin Lymphoma. This structure means that response is potentially recorded for all types of Non-Hodgkin Lymphoma irrespective of the sub-entity to which this case belongs, while staging are recorded differently for each group as is indicated by the inclusion of a staging data item in the specific sub-entity. The diagram also indicates that all forms of cancer may have cytological information recorded and dated and that this information relates specifically to that instance of cancer.

Table 7: Haematological Cancer Groups and Sub-Groups

Haematological Cancer Group Code	Sub-Group	Haematological Cancer Group Name	Primary Site of Cancer (ICD10-AM)	Morphology (ICD-O)
01		Hodgkin Lymphoma	C81 – C81.99	9650/3 – 9659/3
02		Non Hodgkin Lymphoma	C82.0 – C85.9	Any cases in sub-groups 1-3
02	1	Follicular (nodular) Non-Hodgkin Lymphoma	C82.0 – C82.9	9690/3 9691/3 9695/3 9698/3
02	2	Aggressive Non-Hodgkin Lymphoma	C82.0 – C85.9	9680/3 9673/3 9679/3 9680/3 9678/3 9705/3 9702/3 9719/3 9716/3 9714/3
02	8	Other Non-Hodgkin Lymphoma	C82.0 – C85.9	Any not listed elsewhere
03		Multiple Myeloma	C90.0 – C90.01	9732/3
10		Acute Lymphoblastic Leukaemia	C91.0 - C91.01	Any not listed elsewhere
11		Chronic Lymphocytic Leukaemia	C91.1 - C91.11	9823/3
20		Acute Myeloid Leukaemia	C92.0 – C92.01 C92.4 – C92.91 C93.0 – C93.01 C94.0 C94.3 – C94.39 C94.4 – 94.59	9805/3 9840/3 9866/3, 9877/3 9870/3 – 9874/3 9891/3 9895/3 – 9897/3 9910/3 – 9931/3
21		Chronic Myeloid Leukaemia	C92.1 - C92.11	9863/3
30		Myelodysplastic Syndrome	D46.0 - D46.9	9980/3 – 9989/3
80		Other Haematological Cancer	C88.0 – C89.9 C90.1 - C90.29 C92.3 - C96.9	Any not listed elsewhere

Figure 1: Haematological Cancer Data Structure



Worked Example

Example 1:

Mary Smith, Age 45, easy bruising and tiredness
Marrow aspirate shows APML confirmed on FISH for t(15;17).
Therapy on APML4 study protocol
Morphological CR after induction.
Cytogenetic CR confirmed after first consolidation
Molecular CR confirmed after second consolidation
Continuing molecular CR confirmed 6 months later

Data Recorded:

Date of diagnosis of primary Cancer: 1 Jan 05
Primary Site of Cancer: C94.20 Acute Promyelocytic Leukaemia
Morphology of Cancer: 9866/3: Acute Promyelocytic Leukaemia
These data put the cancer into the Acute Myeloid Leukaemia Group to which response, cytogenetic and molecular markers and dates can be added at any time.

Response: 01 - CR: Date: 29 Jan 05 (complete remission)
Response: 33 - CMR: Date: 7 Mar 05 (complete molecular response)
Response: 33 - CMR: Date: 9 Sep 05 (complete molecular response)

Note: demographics, provider details, treatment detail and dates, Clinical trial enrolment and ECOG are specified and recorded according the Clinical Cancer Registry minimum data set.

Example 2:

John Luck, aged 63, tired
Hb 95 g/L, neutrophils $1.2 \times 10^9/L$, platelets $115 \times 10^9/L$
Marrow biopsy consistent with myelodysplasia (RCMD)
Cytogenetics abnormal with 7q- and additional complex abnormalities
Progression to AML 7 months later
Death after a further 3 months having failed induction chemotherapy.

Data Recorded:

Date of diagnosis of Primary Cancer: 1 Jan 05
Primary Site of Cancer: D46.7 Other Myelodysplastic Syndrome
Morphology of Cancer: 9985/3: Refractory cytopenia with multilineage dysplasia
These data put the cancer into the Myelodysplastic Syndrome Group to which prognostic score and cytogenetic and molecular markers and dates can be added at any time.

Prognostic Score: IPSS Score: 1.5 Date: 1 Jan 05

Cytogenetic & Molecular markers:

Marker Type 06 Chromosome 7 abnormality:
Marker Value: +ve - Pos
Date of Report: 1.Jan.05

Cytogenetic & Molecular markers:

Marker Type: 05 - Complex karyotype:
Marker Value: +ve - Pos
Date of Report: 1.Jan.05

Response: 51 - PD: Date: 1 Aug 05 (progressive disease)

Second Diagnosis : 1 Aug 05
Acute Myeloid Leukaemia with multilineage dysplasia (9895/3)

Response: 61 - FR: Date: 1 Sep 05 (failure to respond)
Death recorded in MDS: 1 Nov 05

Example 3:

Norm Easy, Aged 71, lump in left axilla, weight loss and sweats for 3 months.
LD 486 iu/L, ECOG 1
Lymph node biopsy: diffuse large B cell Lymphoma, 1 Jan 05
CT demonstrated lymphadenopathy in mediastinum, para-aortic nodes and a single extranodal mass in the pelvis.
RCHOP chemotherapy achieved CR after 4 cycles.
CR confirmed on CT 6 months later.

Data Recorded:

Date of diagnosis of primary cancer: 1 Jan 05

Primary Site of Cancer: C83.3 Lymphoma – diffuse large B cell

Morphology of Cancer: 9680/3: Diffuse Large B cell Lymphoma

These data put the cancer into the Non-Hodgkin Lymphoma Group and the sub-group - Aggressive Non-Hodgkin Lymphoma to which an Ann Arbor Staging classification can be applied as well as a response an international prognostic index score and cytogenetic and molecular markers and dates can be added at any time.

Stage of Disease (Ann Arbor) IIIBE Date: 10 Jan.05

Prognostic Score (IPI): 3 Date: 10 Jan 05

Response: 01 - CR Date: 15 Mar 05 (Complete remission)

Response: 01 - CR Date: 15 Sep 05 (Complete remission)

Data Dictionary

Data Dictionary General Concepts

The dictionary entries included here are described using terms relevant to both the NSW Cancer data collection and consistent with the Australian Institute of Health and Welfare's METeOR metadata register which now holds the national health data dictionary. This layout has been used to enhance consistency of the definitions over time.

The data items and their description are shown in Table 15

Table 15: Data Item and Descriptions

Item	Description
Data Element Name	The name used in the NSW Cancer data collection.
Synonymous Names	Other names for the same data element such as short names for reporting.
Meteor Name	<p>The data item name suggested for use in the national metadata register. This name has a stylised structure that defines the relationships of the data element. Eg: Haematological Cancer Group has the Meteor name of:</p> <p>Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group.</p> <p>This indicates that the haematological cancer group is a sub-set of a primary site of cancer for a specific individual recorded using ICD-10-AM</p>
Definition	The definition of the data item.
Data Element Concept	The Meteor name of the data element concept (without specifying the value domain to be used)
Object	The 'owner' or object of the item. In the case of haematological cancer group the object is the primary site of the cancer from which the haematological group is determined.
Property	The actual definition of Haematological Cancer Groups value domain
Value Domain	The set of values to be used in describing and representing a data element. A given value domain may be used by more than one data element.
Supplementary Codes	Codes that represent the unknown or unspecified concepts in the value domain.
Specialisation	This indicates that this data element is established as subset of an existing data element. It is not a repetition of the item. The group is automatically established using the values in the original data element to establish the group into which each individual entry falls (the specialisation criteria).

Haematological Cancer Group – Cancer Diagnosis

The Grouping of haematological cancer, particularly the identification of specific groups and the criteria used to define a group must be reviewed by clinical experts to ensure that they are appropriate.

Data Element Name	Haematological Cancer Group
Synonymous Names	WHO codes Cancer Diagnosis
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3 rd edn) ANN{.N[N]}- haematological cancer group
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3 rd edn) ANN{.N[N]}- haematological cancer group
Definition	The haematological malignancy group into which this cancer falls based upon the ICD10-AM code used for the cancer diagnosis (primary site of cancer) for an individual and the morphology of that cancer.
Specialisation	This grouping system is a specialization of the primary site of cancer data element defined by the ICD10-AM code held in that field according to the guide for use.
Object Class	Person with cancer—primary site of cancer, code (ICD–10–AM 3 rd edn) ANN{.N[N]}
Property	Haematological Cancer Group
Guide for Use	Patterns of care studies collect a broader and finer level of information on patients treated within given disease groups. Typically, they assess the compliance of a cohort of cancer patient treatments against best practice guidelines.

This data item is provided to allow the application of a succinct, critical set of indicators for a specific haematological cancer diagnosis group regularly, rather than sporadically.

Groups are allocated according to the values of Person with cancer – primary site of cancer, and Person with cancer morphology according to the table below:

Haematological Cancer Diagnosis Group Code	Haematological Cancer Diagnosis Group Name	Primary Site of Cancer (ICD10-AM)
01	Hodgkin Lymphoma	C81 – C81.99
02	Non Hodgkin Lymphoma	C82.0 – C85.9
03	Multiple Myeloma	C90.0–C90.01
10	Acute Lymphoblastic Leukaemia	C91.0-C91.01
11	Chronic Lymphocytic Leukaemia	C91.1-C91.11
20	Acute Myeloid Leukaemia	C92.0 – C92.01 C92.4 – C92.91 C93.0 – C93.01 C94.0 C94.3 – C94.39 C94.4 – 94.59
21	Chronic Myeloid Leukaemia	C92.1 - C92.11
30	Myelodysplastic Syndrome	D46.0 - D46.9
80	Other Haematological Cancer	C88.0 – C89.9 C90.1 - C90.29 C92.3 - C96.9

Collection Methods

This item can be manually or automatically calculated by identifying the ICD10-AM code applicable to the patient's primary site of cancer and the code values are indicated within the range specified in the permissible values section of the value domain.

Related Metadata References

Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005

Value Domain Definition

Haematological Cancer Group
Code used to represent the haematological cancer group into which the primary cancer of a person with cancer falls.

Representational Class

Code

Data Type Format

Number
NN

Maximum Length

2

Permissible Values

Range of Values	Meaning
01	Hodgkin Lymphoma
02	Non-Hodgkin Lymphoma
03	Multiple Myeloma
10	Acute Lymphoblastic Leukaemia
11	Chronic Lymphocytic Leukaemia
20	Acute Myeloid Leukaemia
21	Chronic Myeloid Leukaemia
30	Myelodysplastic Syndrome
80	Other Haematological Cancer
99	Not applicable

Supplementary Codes

Haematological Cancer Sub-Group

The Grouping of haematological cancer, particularly the identification of specific groups and the criteria used to define a group must be reviewed by clinical experts to ensure that they are appropriate.

Data Element Name	Haematological Cancer Sub-Group
Synonymous Names	
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group – Haematological Sub-Group
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group – sub-group
Definition	Haematological Cancer Groups have been divided for reporting and management purposes. This field identifies the sub group to which this instance of disease belongs.
Object Class	Person with cancer – Haematological Cancer Group
Property	Sub-Group
Guide for Use	

Haematological Cancer Diagnosis Group Code	Sub-Group	Haematological Cancer Diagnosis Group Name	Primary Site of Cancer (ICD10-AM)	Morphology (ICD-O)
02	1	Follicular (nodular) Non-Hodgkin Lymphoma	C82.0 – C82.9	9690/3 9691/3 9695/3 9698/3
02	2	Aggressive Non-Hodgkin Lymphoma	C82.0 – C85.9	9680/3 9673/3 9679/3 9680/3 9678/3 9705/3 9702/3 9719/3 9716/3 9714/3
02	8	Other Non-Hodgkin Lymphoma	C82.0 – C85.9	Any not listed elsewhere

Collection Methods	This item can be manually or automatically calculated by identifying the ICD10-AM code applicable to the patient's primary site of cancer and the morphology relevant to the cancer code values are indicated within the range specified in the permissible values section of the value domain.
Related Metadata	Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005
References	Sub-Group
Value Domain	Code used to represent the haematological cancer sub-group into which the primary cancer of a person with cancer falls.
Definition	Code
Representational Class	Code
Data Type	Number
Format	N

Maximum Length	1	
Permissible Values	Range of Values	Meaning
	1	Follicular (Nodular) Non-Hodgkin Lymphoma
	2	Aggressive Non-Hodgkin Lymphoma
	8	Other Non-Hodgkin Lymphoma
Supplementary Codes	9	Not applicable
Comments	Aggressive lymphomas defined according to the Australian Cancer Network guidelines.	
Related Metadata	Australian Cancer Network Diagnosis and Management of Lymphoma Guidelines Working Party. Guidelines for the Diagnosis and Management of Lymphoma. The Cancer Council Australia and Australian Cancer Network, Sydney 2005.:217	
References		

Prognostic Marker Type

Value Domain	Prognostic Marker Type																																				
Definition	Code used to indicate the type of cytogenetic, molecular or other biological abnormality detected in a person with haematological cancer																																				
Representational Class	Code																																				
Data Type	Numeric																																				
Format	NN																																				
Maximum Length	2																																				
Permissible Values	<table border="0"> <thead> <tr> <th>Value</th> <th>Meaning</th> </tr> </thead> <tbody> <tr><td>01</td><td>t(4:14)</td></tr> <tr><td>02</td><td>t(14:16)</td></tr> <tr><td>03</td><td>Del(5q)</td></tr> <tr><td>04</td><td>Del(20q)</td></tr> <tr><td>05</td><td>Complex Karyotype</td></tr> <tr><td>06</td><td>Chromosome 7 abnormality</td></tr> <tr><td>11</td><td>Del(13q)</td></tr> <tr><td>12</td><td>Del(17p)</td></tr> <tr><td>13</td><td>+8</td></tr> <tr><td>14</td><td>+21</td></tr> <tr><td>15</td><td>+22</td></tr> <tr><td>16</td><td>11q23 abnormality</td></tr> <tr><td>17</td><td>-5</td></tr> <tr><td>18</td><td>-7</td></tr> <tr><td>19</td><td>3q abnormality</td></tr> <tr><td>20</td><td>ZAP-70 overexpression</td></tr> <tr><td>21</td><td>FLT3 mutation</td></tr> </tbody> </table>	Value	Meaning	01	t(4:14)	02	t(14:16)	03	Del(5q)	04	Del(20q)	05	Complex Karyotype	06	Chromosome 7 abnormality	11	Del(13q)	12	Del(17p)	13	+8	14	+21	15	+22	16	11q23 abnormality	17	-5	18	-7	19	3q abnormality	20	ZAP-70 overexpression	21	FLT3 mutation
Value	Meaning																																				
01	t(4:14)																																				
02	t(14:16)																																				
03	Del(5q)																																				
04	Del(20q)																																				
05	Complex Karyotype																																				
06	Chromosome 7 abnormality																																				
11	Del(13q)																																				
12	Del(17p)																																				
13	+8																																				
14	+21																																				
15	+22																																				
16	11q23 abnormality																																				
17	-5																																				
18	-7																																				
19	3q abnormality																																				
20	ZAP-70 overexpression																																				
21	FLT3 mutation																																				
Supplementary Classification																																					
Guide for Use	<p>Multiple Myeloma (haematological cancer group)</p> <ul style="list-style-type: none"> Using interphase FISH, deletion 13q14 and deletion 17p13 were associated with poor response to induction treatment and short median OS. 11q abnormalities also correlated with short median OS. Using interphase FISH, t(4:14)(p16;q32), t(14;16)(q32;q23), deletion 17p13, and deletion 13q14 were associated with shorter survival. <p>Acute Myeloid Leukaemia (haematological cancer group)</p> <ul style="list-style-type: none"> Two specific FLT3 mutations have been described, both associated with decreased survival and increased risk of relapse. One is an internal tandem repeat mutation, the other a missense mutation (D835). Both activate the FLT3 protein. <p>Cytogenetic structural abnormalities, when present, are strongly predictive of outcome in AML.</p>																																				
Collection Methods	Clinician determines status according to instructions in the guide for use.																																				
Comments	Permissible values indicated above are indicative only, pending discussion with relevant clinical experts.																																				
Related Metadata	Is formed using Person with cancer—primary site of cancer,																																				
References	code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.																																				

Konigsberg R, Zojer N, Ackermann J et al. Predictive role of

interphase cytogenetics for survival of patients with multiple myeloma. *Journal of Clinical Oncology* 18: 804-812. 2000.

Fonseca R, Blood E, Montserrat R et al. Clinical and biologic implications of recurrent genomic aberrations in myeloma. *Blood* 101: 4569-4575. 2003.

Grimwade D, Walker H, Oliver F et al. The Importance of Diagnostic Cytogenetics on Outcome in AML: Analysis of 1,612 Patients Entered Into the MRC AML 10 Trial. *Blood*, 92, (7): pp 2322-2333, 1998.

Prognostic Marker Value

This table relates directly to the marker type and is also under review.

Value Domain	Prognostic Marker Value	
Definition	Code used to indicate the value of the cytogenetic abnormality detected in a person with haematological cancer	
Representational Class	Code	
Data Type	Numeric	
Format	N	
Maximum Length	1	
Permissible Values	Value	Meaning
	1	Positive
	0	Negative
	9	Unknown or not recorded
Supplementary Classification		
Guide for Use	Used to record the value of the marker or cytogenetic attribute for this person on this occasion. This item is recorded along with the date recorded, and the marker type.	

Date of Reporting

Synonymous Names	Response date Marker date Prognosis as at date
Meteor Name	Haematological cancer group–date of reporting
Data Element Concept	Haematological Cancer Group – date of reporting
Definition	The date upon which a set of haematological cancer data were reported.
Object Class	Haematological Cancer Group
Guide for Use	Used to indicate the date applicable to a set of details such as staging, prognosis, response, failure, or markers.
Collection Methods	
Value Domain	Date of Reporting
Definition:	Date upon which data was reported
Representational Class	Date
Data Type	Date
Format	DD/MM/YYYY
Maximum Length	10
Permissible Values	Any date in the past

Value Domains

The following value domains are used by more than one of the cancer groups and are therefore shown here in detail rather than repeat them throughout the document.

Stage of Disease (Ann Arbor)

Used for Hodgkin Lymphoma and Non-Hodgkin Lymphoma

Value Domain	Stage of disease (Ann Arbor)	
Definition	The code used to indicate the stage to which Non-Hodgkin Lymphoma or Hodgkin Lymphoma has progressed at the time recorded.	
Classification Scheme	Ann Arbor Staging System for Hodgkin Lymphoma and Non-Hodgkin Lymphoma	
Representational Class	Code	
Data Type	Numeric	
Format	NN	
Maximum Length	2	
Permissible Values	Value	Meaning
	1	IA
	2	IAE
	3	IB
	3	IBE
	11	IIA
	12	IIAE
	13	IIB
	14	IIBE
	21	IIIA
	22	IIIAE
	23	IIIAES
	24	IIIB
	25	IIIBE
	26	IIIBES
	31	IVA
	32	IVB
Reference	Rosenberg SA. <i>Validity of the Ann Arbor staging classification for the non-Hodgkins' lymphomas</i> . Cancer Treat Rev 61 1023-27 (1977)	

Stage of Disease (Binet)

Value Domain	Stage of disease (Binet)
Definition	The code used to indicate the stage of a case of Chronic Lymphocytic Leukaemia at the time recorded.
Classification Scheme	Binet Staging System for Chronic Lymphocytic Leukaemia.
Representational class	Code
Data type	String
Format	A
Maximum Length	1
Permissible Values	A,B,C

Stage of Disease (Durie and Salmon)

Value Domain	Stage of disease (Durie and Salmon)
Definition	The code used to indicate the stage to which this case of multiple myeloma has progressed at the time recorded.
Classification Scheme	Durie & Salmon for Multiple Myeloma
Representational Class	Code
Data Type	Numeric
Format	N
Maximum Length	1
Permissible Values	Value Meaning 1 Stage I 2 Stage II 3 Stage III 9 Unknown
Supplementary Classification	

Stage of Disease (CML)

Value Domain	Stage of disease (CML)
Definition	The code used to indicate the stage to which this case of chronic myeloid leukaemia has progressed at the time recorded.
Classification Scheme	CML staging system
Representational Class	Code
Data type	Numeric
Format	N
Maximum Length	1
Permissible Values	Value Meaning 1 CP – chronic phase 2 AP – accelerated phase 3 BC – blast crisis 9 Unknown
Supplementary Classification	

Prognostic Indicator Score (Sokal)

Used for Chronic Myeloid Leukaemia

Value Domain	Stage of disease (Sokal)
Definition	The code used to indicate the stage to which this case of chronic myeloid leukaemia has progressed at the time recorded.
Classification Scheme	Sokal classification schemes
Representational Class	Code
Data Type	Number
Format	N.NN
Maximum Length	4
Permissible Values	0.01 to 9.99
Supplementary Classification	0.00

Stage of Disease (Rai)

Used for Chronic Lymphocytic Leukaemia

Value Domain	Stage of disease (Rai)												
Definition	The code used to indicate the stage to which chronic lymphocytic leukaemia has progressed at the time of diagnosis.												
Classification Scheme	Rai staging system												
Representational Class	Code												
Data Type	Numeric												
Format	N												
Maximum Length	1												
Permissible Values	<table><thead><tr><th>Value</th><th>Meaning</th></tr></thead><tbody><tr><td>0</td><td>0</td></tr><tr><td>1</td><td>I</td></tr><tr><td>2</td><td>II</td></tr><tr><td>3</td><td>III</td></tr><tr><td>4</td><td>IV</td></tr></tbody></table>	Value	Meaning	0	0	1	I	2	II	3	III	4	IV
Value	Meaning												
0	0												
1	I												
2	II												
3	III												
4	IV												
Supplementary Classification	<table><tbody><tr><td>8</td><td>Not applicable</td></tr><tr><td>9</td><td>Unknown</td></tr></tbody></table>	8	Not applicable	9	Unknown								
8	Not applicable												
9	Unknown												

Hasenclever Prognostic Scoring System Score

These values are used to indicate the score that represents the prognostic estimate at a given date for a case of advanced Hodgkin Lymphoma.

Value Domain	Prognostic Score (Hasenclever)	
Definition	The score achieved for this instance of advanced Hodgkin Lymphoma for this patient on the associated date	
Representational Class	Code	
Data Type	Number	
Format	N	
Maximum Length	1	
Permissible Values	Value	
	0	
	1	
	2	
	3	
	4	
	5	
	6	
	7	
Supplementary Classification	9	Unknown, unspecified

International Staging System Score

These values are used to indicate the score that represents the prognostic estimate at a given date for a case of multiple myeloma.

Value Domain	Prognostic score (ISS)	
Definition	The score achieved for this instance of multiple myeloma for this patient on the associated date	
Representational Class	Code	
Data Type	Number	
Format	N	
Maximum Length	1	
Permissible Values	Value	Meaning
	1	I
	2	II
	3	III
Supplementary Classification	9	Unknown

International Prognostic Scoring System Score

These values are used to indicate the score that represents the prognostic estimate at a given date for a case of myelodysplastic syndrome.

Value Domain	Prognostic Score (IPSS)	
Definition	The score achieved for this instance of myelodysplastic syndrome for this patient on the associated date	
Representational Class	Code	
Data Type	Number	
Format	N.N	
Maximum Length	3	
Permissible Values	Value	Meaning
	0.0	Low
	0.5	Intermediate 1
	1.0	Intermediate 1
	1.5	Intermediate 2
	2.0	Intermediate 2
	2.5	High
	3.0	High
	3.5	High
Supplementary Classification	9.9	Unknown, unspecified

International Prognostic Index Score

These values are used to indicate the score that represents the prognostic estimate at a given date for a case of aggressive Non Hodgkin Lymphoma.

Value Domain	International Prognostic Index Score	
Definition:	The score achieved for this instance of aggressive lymphoma for this patient using the international prognostic index	
Representational Class	Code	
Data Type	Number	
Format	N	
Maximum Length	1	
Permissible Values	Value	Meaning
	0	Low Risk
	1	Low Risk
	2	Low Intermediate Risk
	3	High Intermediate Risk
	4	High Risk
	5	High Risk
	9	Unknown

International Prognostic Index Score – Age Adjusted

These values are used to indicate the score that represents the age adjusted prognostic estimate at a given date for a case of aggressive Non Hodgkin Lymphoma.

Value Domain	International Prognostic Index Score – Age Adjusted	
Definition:	The score achieved for this instance of aggressive lymphoma for this patient using the age adjusted international prognostic index for patients aged less than 61 years.	
Representational Class	Code	
Data Type	Number	
Format	N	
Maximum Length	1	
Permissible Values	Value	Meaning
	0	Low Risk
	1	Low Intermediate Risk
	2	High Intermediate Risk
	3	High Risk
	9	Unknown

Follicular Lymphoma International Prognostic Index Score

Value Domain	Follicular Lymphoma International Prognostic Index Elements	
Definition:	The score achieved for this instance of follicular lymphoma for this patient using the follicular lymphoma international prognostic index	
Representational Class	Code	
Data Type	Number	
Format	N	
Maximum Length	1	
Permissible Values	Value	Meaning
	0	Low
	1	Low
	2	Intermediate
	3	High Intermediate
	4	High
	5	High

Cytogenetic Risk Group

Value Domain	Cytogenetic Risk Group for Acute Myeloid Leukaemia		
Definition:	A cytogenetic risk group determined at diagnosis for cases of Acute Myeloid Leukaemia, determined from bone marrow cytogenetic analysis.		
Representational class	Code		
Data Type	Number		
Format	N		
Maximum Length	1		
Permissible Values	Value	Meaning	
	1	Favourable	
	2	Intermediate	
	3	Adverse	

Response

Value Domain	Response		
Definition	Code used to indicates the level of response to treatment of a haematological cancer		
Representational Class	Code		
Data Type	Number		
Format	NN		
Maximum Length	2		
Permissible Values	Value	Meaning	Alternate Code
	01	Complete Remission	CR
	02	Complete Remission (uncertain)	CRu
	03	Partial Remission	PR
	05	Very Good Partial Response	VGPR
	06	Complete Remission, Incomplete Recovery	CRI
	09	Stringent Complete Remission	sCR
	10	Complete Haematological Response	CHR
	11	Marrow Complete Remission	MaCR
	30	Major Cytogenetic Response	MCyR
	31	Complete Cytogenetic Response	CCyR
	32	Major Molecular Response	MMR
	33	Complete Molecular Response	CMR
	34	Partial Cytogenetic Remission	PcyR
	41	Haematologic Improvement - Erythroid	HI-E
	42	Haematologic Improvement - Platelet	HI-P
	43	Haematologic Improvement - Neutrophil	HI-N
	50	Stable Disease	SD
	51	Progressive Disease	PD
	52	Relapse	RL
	53	Molecular Relapse	MR
	54	Cytogenetic Relapse	CyR
	55	Relapse after Haematologic Improvement	HI-RL
	60	Resistant Disease	RD
	61	Treatment Failure – Indeterminate Cause	TFI
	62	Aplasia	AP
	65	No Response	NR
Supplementary Classification	99	Unknown or not entered or not applicable	

Hodgkin Lymphoma

Hodgkin Lymphoma	
PK	<u>Cancer Diagnosis</u>
	Stage of Disease (Ann Arbor) Prognostic Score (Hasenclever) Response Date of report

Figure 2: Hodgkin Lymphoma Structure

Hodgkin Lymphoma – Stage of Disease (Ann Arbor)

Metadata Type	Data Element
Synonymous Names	Ann Arbor staging of Hodgkin Lymphoma and Non-Hodgkin Lymphoma Ann Arbor Staging Classification
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Hodgkin Lymphoma) – stage of disease (Ann Arbor)
Definition	Describes the stage used in expressing the extent of a specific type of Hodgkin Lymphoma cancer at the time of diagnosis.
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Hodgkin Lymphoma) – stage of disease (Ann Arbor)
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Hodgkin Lymphoma)
Value Domain	Stage of disease (Ann Arbor)
Guide for Use	The Ann Arbor system of staging was developed to characterise the extent of disease in patients with Hodgkin Lymphoma (HL).

The Ann Arbor system was modified at the Cotswolds meeting to include definitions of bulky disease in the CT era (>10 cm), definition of CT criteria for splenic and liver involvement (focal defects), and definition of a new category of treatment response (CR(u)) with persistent radiological abnormalities of uncertain significance.

Recommended staging procedures in Hodgkin Lymphoma:
CT Neck, Chest, Abdomen, Pelvis
Bone marrow biopsy in at least those with stage >IIA or abnormal blood count.
Functional scanning by FDG-PET (Gallium if PET unavailable)

Collection Methods
Comments

Staging systems seek to classify patients having a similar prognosis into groups or stages.

Stage	Criteria
I	Involvement of a single lymph node (I) or of a single extralymphatic organ or site (IE)
II	Involvement of two or more lymph node regions, same side of the diaphragm alone (I) or with involvement of limited, contiguous extralymphatic organ or site (IIE)
III	Involvement of lymph node regions both sides of the diaphragm (III) which may include the spleen (IIIS) and/or limited, contiguous extralymphatic organ or site (IIIE, IIIES)
IV	Multiple or disseminated foci of involvement of one or more extralymphatic organs or tissues, with or without lymphatic involvement
All stages plus A or B	Asymptomatic: (A) With weight loss (10% over 6 months) and/or fever (38°) and/or sweats (drenching, often nocturnal): (B)
E	An E added after the stage number indicates that extranodal involvement is present
S	An S added after the stage number (for stage III) indicates splenic involvement.

**Related Metadata
References**

Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.

Australian Cancer Network Diagnosis and Management of Lymphoma Guidelines Working Party. Guidelines for the Diagnosis and Management of Lymphoma. The Cancer Council Australia and Australian Cancer Network, Sydney 2005.:150.

Hodgkin Lymphoma – Prognostic Score (Hasenclever)

Metadata Type	Data Element
Synonymous Names	Hasenclever Score
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Hodgkin Lymphoma) – Prognostic Score (Hasenclever)
Definition	Describes the likely prognostic risk of this person’s Hodgkin Lymphoma.
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Hodgkin Lymphoma) – Prognostic Score(Hasenclever)
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Hodgkin Lymphoma)
Value Domain	Prognostic Score (Hasenclever)
Guide for Use	For the Hasenclever score, the value domain is specified as values between 0 and 7. The score is calculated by enumerating which of the following conditions are present at diagnosis.

Factor	Relative Risk
Serum Albumin < 40g/litre	1.49
Haemoglobin < 105g/litre	1.35
Male sex	1.35
Stage IV disease	1.26
Age >= 45 years of age	1.39
White-cell count $\geq 15 \times 10^9$ per litre	1.41
Lymphocyte count < 0.6 by 10^9 per litre	1.38

*Relative risks are for freedom from progression of disease in patients with the factors as compared with those without the factors.

Collection Methods Comments

The Hasenclever score has been validated as follows for advanced Hodgkin Disease.
Rates of Freedom from Progression of Disease and Overall Survival at 5 years According to Individual and Grouped Prognostic Scores

Prognostic Score	Rate of Freedom from Progression	Rate of Overall Survival
0	84%	89%
1	77%	90%
2	67%	81%
3	60%	78%
4	51%	61%
≥ 5	42%	56%

Related Metadata References

Is formed using Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.

Hasenclever, D, Diehl, V, A prognostic score for advanced Hodgkin’s Disease. NEJM 2006; 339 (21): 1506-1514.

Hodgkin Lymphoma – Response

Synonymous names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Hodgkin’s Disease) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Hodgkin Lymphoma) – Response
Definition	Indicate the level of response to treatment of this patient with Hodgkin Lymphoma
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Hodgkin Lymphoma)
Value Domain	Response
Guide for Use	<p>02 - Complete Remission (CR): No clinical, radiological or other evidence of HL although changes due to therapy (eg: radiation fibrosis) may be noted.</p> <p>03 - Complete Remission unconfirmed/uncertain (CR(u)): Residual stable abnormalities of uncertain significance on structural imaging at sites of known involvement after an excellent partial remission. Functional imaging should be negative with no clinical evidence of active disease.</p> <p>11 - Partial Remission (PR): Reduction by at least 50% in the sum of the products of the largest perpendicular diameters of all measurable lesions. Resolution of B symptoms and no new lesions.</p> <p>51 - Progression of Disease (PD): Increase by 25% or greater in the size of at least one measurable lesion, appearance of new lesions or recurrence of B symptoms.</p>
Collection Methods	
Comments	
Related Metadata	
References	<p>Is formed using person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.</p> <p>Australian Cancer Network Diagnosis and Management of Lymphoma Guidelines Working Party. Guidelines for the Diagnosis and Management of Lymphoma. The Cancer Council Australia and Australian Cancer Network, Sydney 2005.:165</p>

Non-Hodgkin Lymphoma

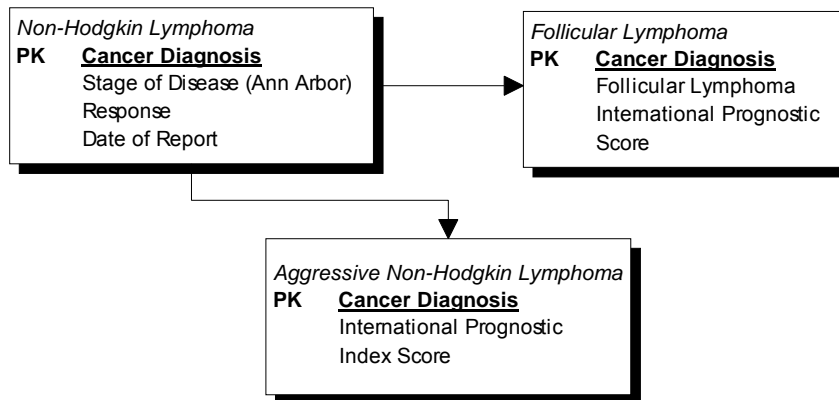


Figure 3: Non-Hodgkin's Lymphoma Structure

All types of Non-Hodgkin Lymphoma collect the data items indicated in the Non-Hodgkin Lymphoma data group. Additional data items are able to be recorded for Follicular Lymphoma and Aggressive Non-Hodgkin Lymphoma.

Non-Hodgkin Lymphoma – Stage of Disease (Ann Arbor)

Metadata Type	Data Element
Synonymous Names	Ann Arbor staging of Hodgkin Lymphoma and Non-Hodgkin Lymphoma Ann Arbor Staging Classification
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Non-Hodgkin Lymphoma) – stage of disease (Ann Arbor)
Definition	Describes the stage used in expressing the extent of a specific type of Non-Hodgkin Lymphoma cancer at the time of diagnosis.
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Non-Hodgkin Lymphoma) – stage of disease (Ann Arbor)
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Hodgkin Lymphoma)
Value Domain	Stage of disease (Ann Arbor)
Guide for Use	<p>The Ann Arbor system of staging was developed to characterise the extent of disease in patients with Hodgkin Lymphoma (HL) but has now been extended and validated in NHL.</p> <p>The Ann Arbor system was modified at the Cotswolds meeting to include definitions of bulky disease in the CT era (>10 cm), definition of CT criteria for splenic and liver involvement (focal defects), and definition of a new category of treatment response (CR(u)) with persistent radiological abnormalities of uncertain significance.</p> <p>Recommended staging procedures in Non-Hodgkin Lymphoma include: CT Chest, Abdomen, Pelvis Bone marrow biopsy Functional scanning by FDG-PET (Gallium if PET unavailable)</p>
Collection Methods	
Comments	Staging systems seek to classify patients having a similar prognosis into groups or stages.

Stage	Criteria
I	Involvement of a single lymph node (I) or of a single extralymphatic organ or site (IE)
II	Involvement of two or more lymph node regions, same side of the diaphragm alone (I) or with involvement of limited, contiguous extralymphatic organ or site (IIE)
III	Involvement of lymph node regions both sides of the diaphragm (III) which may include the spleen (IIIS) and/or limited, contiguous extralymphatic organ or site (IIIE, IIIES)
IV	Multiple or disseminated foci of involvement of one or more extralymphatic organs or tissues, with or without lymphatic involvement
All stages plus A or B	Asymptomatic: (A) With weight loss (10% over 6 months) and/or fever (38 ⁰) and/or sweats (drenching, often nocturnal): (B)
E	An E added after the stage number indicates that extranodal involvement is present
S	An S added after the stage number (for stage III) indicates splenic involvement.

***Related Metadata
References***

Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.

Australian Cancer Network Diagnosis and Management of Lymphoma Guidelines Working Party. Guidelines for the Diagnosis and Management of Lymphoma. The Cancer Council Australia and Australian Cancer Network, Sydney 2005, 218.

Lister TA, Crowther D, Sutcliffe SB et al: Report of a Committee to discuss the Evaluation and Staging of Patients with Hodgkin's Disease: Cotswolds Meeting. J Clin Oncol 7:1630-1636,1989.

Non-Hodgkin Lymphoma – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Non Hodgkin Lymphoma) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Non-Hodgkin Lymphoma) – Response
Definition	Indicate the level of response to treatment of this patient with Non-Hodgkin Lymphoma
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Non-Hodgkin Lymphoma)
Value Domain	Response
Guide for Use	For this disease condition the following criteria are used to indicate response or remission (the IWC criteria):

01 Complete Response or Remission (CR)

- CR based on previous IWC criteria
 1. Complete disappearance of all detectable clinical and radiological evidence of disease and disease related symptoms, normalisation of biochemical abnormalities such as raised LDH (criteria 1)
 2. Lymph nodes and masses must regress to normal size ($\leq 1.5\text{cm}$ for nodes $> 1.5\text{ cm}$ before therapy). Nodes 1.1 to 1.5 cm before treatment must have decreased to $\leq 1\text{cm}$ in their greatest transverse diameter after treatment, or by $> 75\%$ in the sum of the products of the greatest diameters (SPD) (criteria 2)
 3. Regression of previous splenomegaly which is not palpable on clinical examination post therapy (criteria 3)
 4. Clearance of bone marrow involved by lymphoma prior to therapy (criteria 4) AND
- **PET Criteria**
 - CR by IWC and Completely Negative PET
 - CRu, PR or SD by IWC, Completely Negative PET, Negative BMB if positive prior to therapy.
 - PD by IWC with Completely Negative PET, Negative BMB if positive prior to therapy (Only if prior lesion $\geq 1.5\text{cm}$ or 1 cm in lung)

02 Complete Response unconfirmed (CRu)

- Criteria 1 and 3 of complete response with
 - Residual lymph node mass $> 1.5\text{cm}$ in greater transverse diameter that has regressed by more than 75% in the SPD AND/OR
 - Indeterminate bone marrow (increased number of size of aggregates without cytologic or architectural atypia) PLUS
 - Completely negative PET
- **PET Criteria**
 - CRu by IWC, Completely Negative PET, but Indeterminate bone marrow

03 Partial Response (PR)

- $\geq 50\%$ decrease in SPD of the six largest dominant nodes or nodal masses (regions chosen should be as disparate as possible and include mediastinal and retroperitoneal areas if involved)
- No increase in the size of other nodes, liver or spleen
- Splenic and hepatic nodules must regress by $\geq 50\%$ in the SPD
- No new sites of disease PLUS
- Completely negative PET and BM negative if positive prior to therapy.
- **PET Criteria**
 - CR, CRu, PR by IWC and a Positive PET at a site of previous disease.
 - CR, CRu, PR, SD by IWC and a Positive PET outside the site of previous disease.
 - SD by IWC with a Positive PET at a previously involved nodal site that had regressed to $< 1.5\text{cm}$ (if previously $> 1.5\text{cm}$) or $< 1\text{cm}$ (if previously $1.1\text{-}1.5\text{cm}$).

50 Stable Disease (SD)

- Less than a PR but not satisfying the criteria for progressive disease.
- **PET Criteria**
 - SD by IWC with a Positive PET at a previously involved nodal site (ie residual mass)

51 Progressive Disease (PD)

- Progression from PR or non-responders.
- $\geq 50\%$ increase from nadir in SPD of any previously abnormal node.
- Appearance of any new lesion during or after therapy.
- **PET Criteria**
 - PD by IWC and a Positive PET corresponding to the CT finding
 - PD by IWC with a Negative PET and a CT abnormality (new lesion or increasing old lesion) $< 1.5\text{cm}$ or $< 1\text{cm}$ in lung

52 Relapse (RL)

- Progression from CR or Cru
- Appearance of new lesion or increase by $\geq 50\%$ in site of prior involvement.
- $\geq 50\%$ increase in greatest diameter of any node $< 1\text{cm}$ short axis or in the SPD of more than one node.

Collection Methods

Clinician determines status according to instructions in the guide for use.

Comments

This field has an associated date.

***Related Metadata
References***

Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.

Haematological cancer group – response – date recorded

Cheson BD, Horning SJ, Coiffier B et al: Report of an international Workshop to Standardise Response Criteria for Non-Hodgkin's Lymphomas. J Clin Oncol 17: 1244-1253. 1999.

Judweid ME, Wiseman GA, Vose JM et al: Response Assessment of Aggressive Non-Hodgkin's Lymphoma by Integrated International Workshop Criteria and Fluorine-18-Fluorodeoxyglucose Positron Emission Tomography. J Clin Oncol 23:4652-4661, 2005.

Aggressive Non-Hodgkin Lymphoma - International Prognostic Index Score

Synonymous Names	NHL prognostic score IPI Prognostic Score
Definition	Prognostic score from international prognostic index calculation for this patient for this haematological malignancy
Data Element Concept	Person with cancer – primary site of cancer - Haematological Cancer Group(Non-Hodgkin Lymphoma) – sub-group (2) – IPI prognostic score
Definition	The result of the International Prognostic Index calculation for this for Haematological Cancer sub-group
Object Class	Haematological Cancer Group – sub-group
Property	International Prognostic Index Score
Guide for Use	The IPI uses a model of five prognostic indicators, with patients scoring a point depending on presence/absence of these criteria. The total point score is then used to stratify the patient according to risk.

The age adjusted IPI is applicable to patients aged 60 years or younger. It is recommended that the standard IPI is also recorded.

The following criteria score one point:

Age >60
Ann Arbor clinical stage III or IV
Lactate dehydrogenase (LDH) above normal
Number of extranodal sites >1
ECOG ≥ 2 (ECOG 2 is less than 50% time in bed)

Knowledge table example follows:

Score	Risk	Median 5 year survival
0,1	Low	73%
2	Low Intermediate	51%
3	High Intermediate	43%
4,5	High	26%

Note: Above outcomes reflect therapeutic options in 1993 and may not be valid with current therapies

Value Domain
Related Metadata
References:

International Prognostic Index Score
Shipp MA, Harrington DP, Anderson JR, et. al. The International Non-Hodgkin Lymphoma Prognostic Factors Project: A predictive model for aggressive non-Hodgkin Lymphoma. New England Journal of Medicine 329:987-994. 1993

Nicolaidis C, Fountzilas N, Zoumbos N, et. al. Diffuse large cell lymphomas: Identification of prognostic factors and validation of the International Non-Hodgkin Lymphoma Prognostic Index. Oncology 55: 405-415 1998

Aggressive Non-Hodgkin Lymphoma - International Prognostic Index Score – Age Adjusted

Synonymous names	NHL prognostic score Age Adjusted IPI
Definition	Prognostic score from international prognostic index calculation for this patient for this haematological malignancy. Limited to patients aged 60 years or less.
Data Element Concept	Person with cancer – primary site of cancer - Haematological Cancer Group(Non-Hodgkin Lymphoma) – sub-group (2) – IPI – Age Adjusted prognostic score
Definition	The result of the International Prognostic Index calculation for this for Haematological Cancer sub-group
Object Class	Haematological Cancer Group – sub-group
Property	Age Adjusted International Prognostic Index Score
Guide for Use	The age adjusted IPI is only applicable to patients aged ≤60 years at diagnosis.

The Age Adjusted IPI uses a model of three prognostic indicators, with patients scoring a point depending on presence/absence of these criteria. The total point score is then used to stratify the patient according to risk.

The standard IPI is applicable to all patients and should be recorded as well as the age adjusted IPI to allow broader stratification where appropriate.

The following criteria score one point:

Ann Arbor clinical stage III or IV
Lactate dehydrogenase (LDH) above normal
ECOG ≥ 2 (ECOG 2 is less than 50% time in bed)

Score of 0: low risk
Score of 1: low intermediate risk
Score of 2: high intermediate risk
Score of 3: high risk

Knowledge table example follows:

Score	Risk	CR Rate	2 yr Relapse Free Survival	5 yr Relapse Free Survival
0	Low	92%	88%	86%
1	Low Intermediate	78%	74%	66%
2	High Intermediate	57%	62%	53%
3	High	46%	61%	58%

Note: Above outcomes reflect therapeutic options in 1993 and may not be valid with current therapies

Value Domain	Age Adjusted International Prognostic Index Score
Related Metadata	Shipp MA, Harrington DP, Anderson JR, et. al. The International Non-Hodgkin Lymphoma Prognostic Factors Project: A predictive model for aggressive non-Hodgkin Lymphoma. New England Journal of Medicine 329:987-994. 1993.
References	

Follicular Lymphoma – Follicular Lymphoma International Prognostic Index Score

Synonymous Names	FLIPI score FLIPI Prognostic Score
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group – sub-group (Follicular Lymphoma) – follicular lymphoma international prognostic score
Data Element Concept	Person with cancer – primary site of cancer - Haematological Cancer Group(Follicular Lymphoma) – follicular lymphoma International Prognostic score
Definition	The result of the Follicular Lymphoma International Prognostic Index calculation for this for Haematological Cancer
Object Class	Haematological Cancer Group
Property	Follicular Lymphoma International Prognostic Index Elements
Value Domain	Follicular Lymphoma International Prognostic Index score
Guide for Use	The FLIPI uses a model of five prognostic indicators, with patients scoring a point depending on presence/absence of these criteria. The total point score is then used to stratify the patient according to risk. There is also an age adjusted IPI not shown here.

Follicular Lymphoma International Prognostic Index (FLIPI) uses five prognostic factors, with patients scoring a point if they satisfy each criteria

Age >60
Ann Arbor stage III or IV
Number of nodal areas >4
Serum LDH above normal
Hb ≥ 120g/L

The total score is used to stratify risk
Score of 0 or 1: low risk
Score of 2: intermediate risk
Score of 3 or more: poor risk

Collection Methods Once collected the score is used to identify the level of risk and from that the median 5 year survival shown below:

Knowledge table example:

Score	Risk	5 year survival (%)	10 year survival (%)
0,1	Low	90.6	70.7
2	Intermediate	77.6	50.9
3	High	52.5	35.2

Related Metadata References Solal-Celigny P, Roy P, Colombat P, et al. Follicular lymphoma International Prognostic Index. Blood 104:1258-1265. 2004

Acute Myeloid Leukaemia

Acute Myeloid Leukaemia	
PK	Cancer Diagnosis
	Response
	Date of report

Figure 4: Acute Myeloid Leukaemia Structure

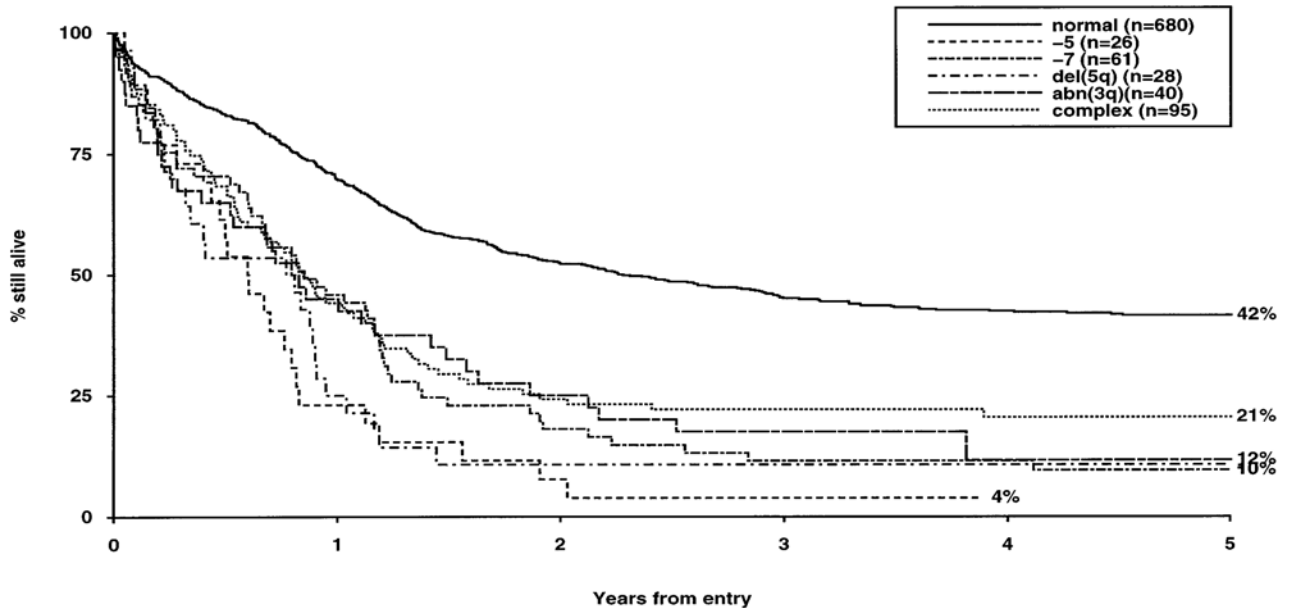
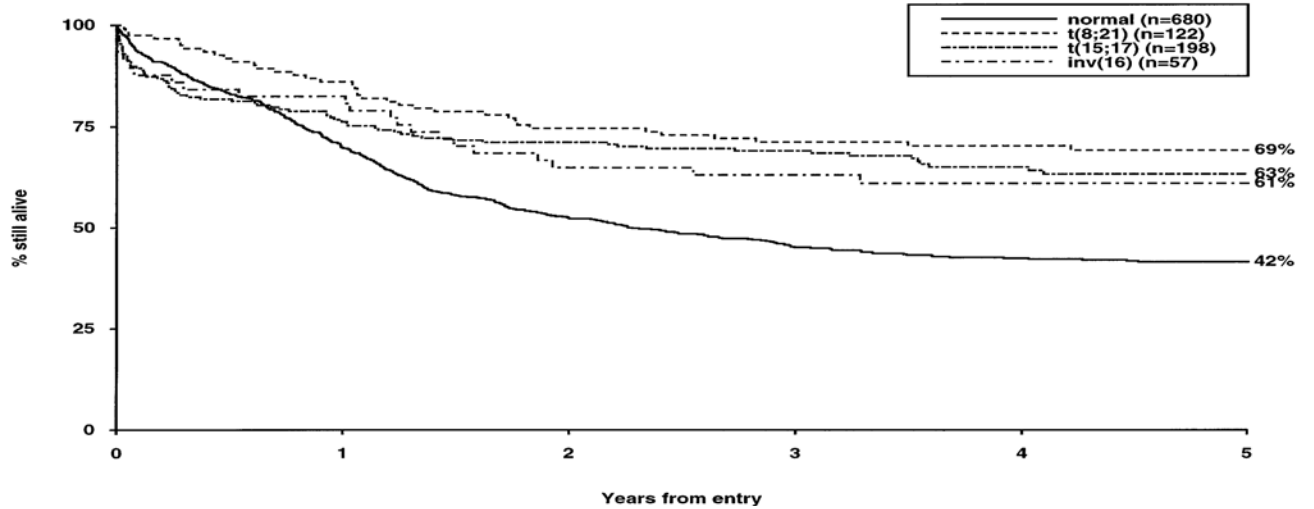
Acute Myeloid Leukaemia – Cytogenetic Risk Group

Synonymous Names	Grimwade prognostic group.
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Acute Myeloid Leukaemia) – Cytogenetic Risk Group
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Acute Myeloid Leukaemia) – Cytogenetic Risk Group
Definition	Indicate the prognostic group based on cytogenetics for this patient with acute myeloid leukaemia.
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Acute Myeloid Leukaemia)
Value Domain	Cytogenetic Risk Group
Guide for use	For this disease condition the following criteria are used to indicate the cytogenetic risk group:

Risk Group	Abnormality	Comment
Favourable	t(8;21) t(15;17) inv(16)	Whether alone or in conjunction with other abnormalities
Intermediate	Normal +8 +21 +22 del(7q) del(9q) Abnormal 11q23 All other structural/ numerical abnormalities	i.e., Cytogenetic abnormalities not classified as favourable or adverse. Lack of additional favourable or adverse cytogenetic changes
Adverse	-5 -7 del(5q) Abnormal 3q Complex	Whether alone or in conjunction with intermediate-risk or other adverse-risk abnormalities

Collection methods

Clinician determines status according to results of bone marrow cytogenetic analysis at diagnosis.



Comments

This field has an associated date.

Overall survival of patients with adverse cytogenetic abnormalities, irrespective of the presence of additional abnormalities. The group with normal karyotype is included for comparison. (Grimwade et al, Blood 92, 2326)

CR Rates, Reasons for Failure, Relapse Risk, and Survival by Hierarchical Cytogenetic Risk Group

Group	No. of Patients	CR (%)	ID (%)	RD (%)	Relapse Risk at5yr % (SE)	Survival at5yr % (SE)
Favorable	377	91*	8	1*	35 (2.8)*	65 (2.5)*
Intermediate	1,072	86	6	8	51 (1.8)	41 (1.5)
Adverse	163	63	14	23	76 (4.5)	14 (2.8)

*P , .001, P values are for Mantel-Haenszel (CR and reasons for failure) or log rank (relapse risk and overall survival) test for trend. (Grimwade et al, Blood 92, 2326)

***Related metadata
references***

Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs. Haematological cancer group – Cytogenetic risk group – date recorded

Grimwade D, Walker H, Oliver F et al. The Importance of Diagnostic Cytogenetics on Outcome in AML: Analysis of 1,612 Patients Entered Into the MRC AML 10 Trial. *Blood*, 92, (7): pp 2322-2333, 1998

Acute Myeloid Leukaemia – Response

Synonymous names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Acute Myeloid Leukaemia) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Acute Myeloid Leukaemia) – Response
Definition	Indicate the level of response to treatment of this patient with acute myeloid leukaemia.
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Acute Myeloid Leukaemia)
Value Domain	Response
Guide for use	For this disease condition the following criteria are used to indicate response or remission:

01 Complete Remission (CR)

IWC Morphologic Complete Remission

- Morphologic leukemia free state: less than 5% blasts on bone marrow aspirate with spicules and with count of at least 200 nucleated cells; no Auer rods, no persisting extramedullary disease, no persistence of unique phenotype by flow cytometry
- Absolute neutrophil count of $>1 \times 10^9/L$
- Platelet count of $100 \times 10^9/L$
- Independent of packed cell transfusions

03 Partial remission (PR)

- Plt count and neutrophil count as for CR
- $\geq 50\%$ reduction in blasts to between 5% and 25% in the bone marrow aspirate
- May require repeat BM after several weeks to distinguish PR from increased blasts caused by BM regeneration
- Generally only relevant in Phase I,II trials

06 Complete Remission, incomplete recovery(CRI)

IWC Morphologic Complete Remission

- Morphologic leukemia free state: less than 5% blasts on bone marrow aspirate with spicules and with count of at least 200 nucleated cells; no Auer rods, no persisting extramedullary disease, no persistence of unique phenotype by flow cytometry
- Neutrophil recovery may be incomplete
- Platelet recovery may be incomplete

31 Complete Cytogenetic Remission (CCyR)

- Criteria for CR met
- Reversion to normal karyotype

33 Complete Molecular Remission (CMR)

- Criteria for CR met
- Failure to detect specific marker present at diagnosis by sensitive molecular method (eg PML/RAR α , AML1/ETO,

CBF β /MYH11)

Treatment Failures are recorded using any of the following codes:

52 Relapse (RL):

- morphological relapse after CR is reappearance of leukemic blasts in peripheral blood or $\geq 5\%$ blasts in BM not attributable to other cause (i.e. BM regeneration)
- also includes new dysplasia or reappearance/development of cytologically proven extramedullary disease

53 Molecular Relapse (MR): reappearance of molecular abnormality, eg: reappearance of PML-RARA fusion transcript on RT-PCR

54 Cytogenetic Relapse (CyR): reappearance of cytogenetic abnormality, eg: reappearance of t(8;21), t(15;17), inv(16)

60 Resistant disease (RD):

- patient survives ≥ 7 days post chemotherapy
- persistent AML IN blood/bone marrow as defined by lack of CR/PR

61 Treatment Failure - Indeterminate Cause (TFI):

- patients who die < 7 days post therapy
- or patients who die > 7 days post therapy with no PB blasts, but no BM
- or patients who did not complete their first course of therapy

62 Aplasia (AP):

- patient survives ≥ 7 days post chemotherapy
- death while cytopenic
- hypoplastic bone marrow within 7 days of death)

Collection Methods

Clinician determines status according to instructions in the guide for use.

Comments

This field has an associated date.

The IWC criteria: 'Early treatment assessment' and 'Morphologic leukemia free state' have not been included at this stage.

**Related Metadata
References**

Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.

Haematological cancer group – response – date recorded

Cheson BD, Bennett JM, Kopecky J et al: Revised Recommendations of the International Working Group for Diagnosis, Standardisation of Response Criteria, Treatment Outcomes and Reporting Standards for Therapeutic Trials in Acute Myeloid Leukemia. JCO 24:4642-4649, 2003.

Chronic Myeloid Leukaemia

Chronic Myeloid Leukaemia	
PK	Cancer Diagnosis
	Stage of Disease (CML) Prognostic Score (Sokal) Response Date of report

Figure 5: Chronic Myeloid Leukaemia

Chronic Myeloid Leukaemia – Stage of Disease (CML)

Metadata Type	Data Element
Synonymous Names	CML Stage
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Myeloid Leukaemia_ – stage of disease (CML)
Definition	Describes the stage used in expressing the extent of a specific type of Chronic Myeloid Leukaemia.
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Non-Hodgkin Lymphoma) – stage of disease (CML)
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Chronic Myeloid Leukaemia)
Value Domain	Stage of disease (CML)
Guide for Use	<p>1 Chronic Phase (CP): Neither accelerated phase or blast crisis.</p> <p>2 Accelerated Phase (AP): One or more of the following: Blasts 10-19% of peripheral blood leukocytes or of nucleated bone marrow cells. Peripheral blood basophils $\geq 20\%$ Persistent Thrombocytopenia ($<100 \times 10^9/L$) unrelated to therapy or persistent thrombocytosis ($>1000 \times 10^9/L$) unresponsive to therapy. Increasing spleen size and increasing WBC unresponsive to therapy. Cytogenetic evidence of clonal evolution.</p> <p>3 Blast Crisis (BC): One or more of the following: Blasts $\geq 20\%$ of peripheral blood leukocytes or of nucleated bone marrow cells. Extramedullary blast proliferation. Large foci or clusters of blasts in the marrow biopsy.</p>
Collection Methods	
Comments	<p>Staging systems seek to classify patients having a similar prognosis into groups or stages.</p> <p>The disease may progress and subsequently respond to therapy, resulting in more than one potential entries for stage in a given patient during the course of their leukaemia.</p>
Related Metadata	Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182,
References	01/03/2005 and the haematological cancer group to which this belongs.

Jaffe E. S., Harris N. L., Stein H ., Vardiman J.W., (Eds.): World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Haematopoietic and Lymphoid Tissues IARC Press Lyon 2001: 21-23.

Chronic Myeloid Leukaemia – Prognostic Indicator Score (Sokal)

Metadata Type	Data Element
Synonymous Names	Sokal Index
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Myeloid Leukaemia_ – Prognostic Indicator Score (Sokal)
Definition	Describes the stage used in expressing the extent of a specific type of Chronic Myeloid Leukaemia.
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Non-Hodgkin Lymphoma) – Prognostic Indicator Score (Sokal)
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Chronic Myeloid Leukaemia)
Value Domain	Prognostic Indicator Score (Sokal)
Guide for Use	The Sokal score is calculated as a hazard ratio $= \exp \{0.0116.(age - 43.4) + 0.0345.(spleen - 7.51 \text{ cm}) + 0.188.[(\text{platelets}/700)^2 - 0.563] + 0.0887.(\% \text{ blasts in blood} - 2.1)\}.$ <p>Risk bands as follows: Low: <0.8 Intermediate: 0.8-1.2 (inclusive) High: >1.2</p>
Collection Methods	
Comments	The Sokal Index was validated for ‘good risk’ CML, excluding blast crisis. Information regarding the outcome in each group from the original paper is not currently relevant due to changes in therapy.
Related Metadata	Is formed using Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.
References	Sokal JE, Cox EB, Baccarani M et al: Prognostic Discrimination in “Good Risk” Chronic Granulocytic Leukaemia. Blood 63:789-799, 1984.

Chronic Myeloid Leukaemia – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Myeloid Leukaemia) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Myeloid Leukaemia) – Response
Definition	Indicate the level of response to treatment of this patient with Chronic Myeloid Leukaemia
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Chronic Myeloid Leukaemia)
Value Domain	Response
Guide for Use	For this disease the responses are likely to be prognostic factors.

10 Complete Haematological Response (CHR)

- Total WCC < 10 x 10⁹/L
- Platelet count < 450 x 10⁹/L
- No immature cells (blasts, promyelocytes, myelocytes) in the peripheral blood
- Disappearance of all signs and symptoms relating to leukaemia, including palpable splenomegaly
- Above lasting for at least 4 wks

30 Major Cytogenetic Response (MCR)

- <35% Ph-positive cells on karyotyping

31 Complete Cytogenetic Response (CCR)

- 0% Ph-positive cells on karyotyping

32 Major Molecular Response (MMR)

- BCR-ABL/ABL ratios of <0.05%, approximately equivalent to a >3 log reduction in BCR-ABL transcript levels

33 Complete Molecular Response (CMR)

- Undetectable bcr-abl transcript levels by sensitive Q-RT-PCR on blood/bone marrow aspirate

Collection Methods Clinician determines status according to instructions in the guide for use.

Comments This field has an associated date.

Related Metadata Is formed using Person with cancer—primary site of cancer,

References code (ICD–10–AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.

Haematological cancer group – response – date recorded

Kantarjian H, Cortes J, O'Brien S. Long term survival benefit and improved complete cytogenetic and molecular response rates with imatinib mesylate in Philadelphia chromosome-positive chronic-phase chronic myeloid leukemia after failure of interferon-alfa. *Blood*. 104:1979-1987. 2004.

Acute Lymphoblastic Leukaemia

Acute Lymphoblastic Leukaemia	
PK	Cancer Diagnosis
	Response
	Date of report

Figure 6: Acute Lymphoblastic Leukaemia Structure

A specific stage of disease classification has not been determined for this cancer type. If a stage is defined, put the value into the appropriate data item indicating thereby the classification system used.

Acute Lymphoblastic Leukaemia – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Acute Lymphoblastic Leukaemia) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Acute Lymphoblastic Leukaemia) – Response
Definition	Indicate the level of response to treatment of this patient with Acute Lymphoblastic Leukaemia
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Acute Lymphoblastic Leukaemia)
Value Domain	Response
Guide for Use	For this disease condition the following criteria are used to indicate response or remission: <p>01 Complete Remission (CR):</p> <ul style="list-style-type: none"> • Absence of symptoms and signs of leukaemia • Neutrophils > 1.0 x10⁹/L • Platelets > 100 x10⁹/L • No leukaemic cells in peripheral blood • Normocellular bone marrow with less than 5% blasts.
Collection Methods	Clinician determines status according to instructions in the guide for use.
Comments	This field has an associated date.
Related Metadata	Is formed using Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.
References	Haematological cancer group – response – date recorded Response criteria from ALLG ALL3 protocol.

Chronic Lymphocytic Leukaemia

Chronic Lymphocytic Leukaemia	
PK	Cancer Diagnosis
	Stage of Disease (Rai)
	Response
	Date of report

Figure 7: Chronic Lymphocytic Leukaemia Structure

A specific stage of disease classification has not been determined for this cancer type. If a stage is defined, put the value into the appropriate data item indicating thereby the classification system used.

Chronic Lymphocytic Leukaemia – Stage of Disease (Rai)

Metadata Type	Data Element												
Synonymous Names	Rai Staging Classification												
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Lymphocytic Leukaemia – stage of disease (Rai)												
Definition	Describes the stage used in expressing the extent of a specific case of chronic lymphocytic leukaemia												
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Lymphocytic Leukaemia) – stage of disease (Rai)												
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Chronic Lymphocytic Leukaemia)												
Value Domain	Stage of disease (Rai)												
Guide for Use	<table> <thead> <tr> <th>Value</th> <th>Meaning</th> </tr> </thead> <tbody> <tr> <td>0</td> <td>0 Lymphocytosis in blood or bone marrow</td> </tr> <tr> <td>1</td> <td>I Lymphocytosis + enlarged lymph nodes</td> </tr> <tr> <td>2</td> <td>II Lymphocytosis + enlarged liver or spleen with or without lymphadenopathy</td> </tr> <tr> <td>3</td> <td>III Lymphocytosis + anaemia (Hb <110 g/L) with or without enlarged liver, spleen, or lymph nodes</td> </tr> <tr> <td>4</td> <td>IV Lymphocytosis + thrombocytopenia (plt count < 100x10⁹/L) with or without anaemia or enlarged liver, spleen or lymph nodes</td> </tr> </tbody> </table>	Value	Meaning	0	0 Lymphocytosis in blood or bone marrow	1	I Lymphocytosis + enlarged lymph nodes	2	II Lymphocytosis + enlarged liver or spleen with or without lymphadenopathy	3	III Lymphocytosis + anaemia (Hb <110 g/L) with or without enlarged liver, spleen, or lymph nodes	4	IV Lymphocytosis + thrombocytopenia (plt count < 100x10 ⁹ /L) with or without anaemia or enlarged liver, spleen or lymph nodes
Value	Meaning												
0	0 Lymphocytosis in blood or bone marrow												
1	I Lymphocytosis + enlarged lymph nodes												
2	II Lymphocytosis + enlarged liver or spleen with or without lymphadenopathy												
3	III Lymphocytosis + anaemia (Hb <110 g/L) with or without enlarged liver, spleen, or lymph nodes												
4	IV Lymphocytosis + thrombocytopenia (plt count < 100x10 ⁹ /L) with or without anaemia or enlarged liver, spleen or lymph nodes												
Collection Methods	Clinician determines status.												
Comments	This field has an associated date.												
References	Rai KR, Sawitsky A, Cronite EP et al. Clinical Staging of Chronic Lymphocytic Leukemia. Blood; 46(2): 219-234; 1975.												

Chronic Lymphocytic Leukaemia – Stage of Disease (Binet)

Metadata Type	Data Element
Synonymous Names	Binet Staging Classification
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Lymphocytic Leukaemia – stage of disease (Binet)
Definition	Describes the stage used in expressing the extent of a specific case of chronic lymphocytic leukaemia
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Lymphocytic Leukaemia) – stage of disease (Binet)
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Chronic Lymphocytic Leukaemia)
Value Domain	Stage of disease (Binet)
Definition	The code used to indicate the stage of a case of Chronic Lymphocytic Leukaemia at the time recorded.
Classification Scheme	Binet Staging System for Chronic Lymphocytic Leukaemia.
Representational Class	Code
Data Type	String
Format	A
Maximum Length	1
Permissible Values	

Values	Lymphadenopathy	Haemoglobin (g/L)	Platelets (x10 ⁹ /L)
A	Less than 3 lymph node groups enlarged	100 or more	100 or more
B	3 or more lymph nodal groups enlarged	100 or more	100 or more
C	As for 'B'	Either <100	Or <100

Comments	This field has an associated date.
Related Metadata	Is formed using Person with cancer—primary site of cancer, code
References	(ICD–10–AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs. Haematological cancer group – response – date recorded Binet JL, Auquier A, Dighiero G et al. A new prognostic classification of chronic lymphocytic leukemia derived from a multivariate survival analysis. <i>Cancer</i> 1981; 48: 198-206.

Chronic Lymphocytic Leukaemia – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Lymphocytic Leukaemia) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group (Chronic Lymphocytic Leukaemia) – Response
Definition	Indicate the level of response to treatment of this patient with Chronic Lymphocytic Leukaemia
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Chronic Lymphocytic Leukaemia)
Value Domain	Response
Guide for Use	<p>01 Complete Remission (CR): Normal physical examination, No symptoms. Lymphocytes $\leq 4 \times 10^9/L$ Neutrophils $\geq 1.5 \times 10^9/L$ Platelets $> 100 \times 10^9/L$ Haemoglobin $> 110 \text{ g/L}$ (untransfused) Bone marrow lymphocytes $< 30\%$ (no nodules) Duration ≥ 2 months</p> <p>03 Partial Remission (PR): Physical examination: $\geq 50\%$ decrease (nodes and/or liver, spleen) Plus ≥ 1 of: Neutrophils $\geq 1.5 \times 10^9/L$ Platelets $> 100 \times 10^9/L$ Haemoglobin $> 110 \text{ g/L}$ (or 50% improvement) Duration ≥ 2 months</p> <p>51 Progressive Disease (PD): Physical examination: $\geq 50\%$ increase or new (nodes and/or liver, spleen) Circulating Lymphocytes $\geq 50\%$ increase Richters Syndrome</p> <p>50 Stable Disease (SD) Not as defined above.</p>
Collection Methods	Clinician determines status according to instructions in the guide for use.
Comments	This field has an associated date.
Related Metadata	Is formed using Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) METeOR identifier 270182, 01/03/2005 and the haematological cancer group to which this belongs.
References	Haematological cancer group – response – date recorded Cheson BD, Bennett JM, Grever M et al. National Cancer Institute-Sponsored Guidelines for Chronic Lymphocytic Leukaemia: Revised Guidelines for Diagnosis and Treatment. Blood 87: 4990-4997, 1996.

Multiple Myeloma

Multiple Myeloma	
PK	Cancer Diagnosis
	Response
	Prognostic Score (ISS)
	Date of report

Figure 8: Multiple Myeloma Structure

Multiple Myeloma – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD–10–AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Multiple Myeloma) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Multiple Myeloma) – Response
Definition	Indicate the level of response to treatment of this patient with Multiple Myeloma
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Multiple Myeloma)
Value Domain	Response
Guide for Use	Parameters utilised: serum EPG and IFE, urine EPG and IFE, bone marrow plasma cell percentage, bone marrow trephine, imaging: lytic lesions, calcium.

01 Complete Response (CR):

- No detectable M-protein in serum and urine by immunofixation.
- No more than than 5% plasma cells in bone marrow.
- Disappearance of soft tissue plasmacytomas.

09 Stringent Complete Remission (sCR)

- CR as defined above.
- Normal free light chain ratio.
- Absence of clonal plasma cells in bone marrow by immunohistochemistry or immunofluorescence.

05 Very Good Partial Response (VGPR):

- Serum and Urine M-protein detectable on Immunofixation but NOT by EPG
- OR**
- $\geq 90\%$ reduction in serum M-protein and urine M-protein <100mg per 24 hours.

03 Partial Response (PR):

- $\geq 50\%$ reduction in serum M-protein.
- Reduction in 24h Urine M-protein excretion by $\geq 90\%$ or to <200mg per 24 hours.
- $\geq 50\%$ reduction in size of soft tissue plasmacytomas, if present.
- **If the serum and urine M-protein are unmeasurable:**
 - $\geq 50\%$ decrease in the difference between involved and

uninvolved Free Light Chain (FLC) levels is required in place of the M-protein criteria.

- ***If the serum and urine M-protein are unmeasurable AND serum free light chains are unmeasurable:***
 - $\geq 50\%$ reduction in marrow plasma cells is required in place of M-protein criteria, provided that baseline marrow plasma cell involvement was no less than 30%.

65 No Response (NR):

- Not meeting criteria for CR, sCR, VGPR or PR

52 Relapse from CR (RL):

- Intended for estimation of disease free survival.
- Indicates relapse from CR or sCR
- Reappearance of Serum or Urine M-protein.
- $\geq 35\%$ Plasma cells in marrow aspirate or biopsy.
- Development of new lytic lesions or development of soft tissue plasmacytoma.
- Hypercalcaemia ($>2.8\text{mmol/L}$) not attributable to other causes.

51 Progressive Disease (PD):

- Intended for estimation of time to progression and progression free survival.
- Applies to primary resistant disease and to patients progressing on or off therapy.
- $>25\%$ increase in serum paraprotein, at least 5g/L.
- $>25\%$ increase in 24h Urine light chain excretion, at least 200mg.
- $>25\%$ increase in plasma cells (at least 10%) in marrow aspirate or biopsy.
- $>25\%$ increase in difference between involved and uninvolved free light chain levels, at least 10mg/dL.
- New bone lesions or soft tissue plasmacytomas
- Increase in size (50% and $\geq 1\text{cm}$) or number of lytic bone lesions. Size measured as sum of products of cross diameters.
- Hypercalcaemia ($>2.8\text{mmol/L}$) not attributable to other causes.
- Decrease in Haemoglobin $\geq 20\text{g/L}$
- Increase in Creatinine $\geq (2\text{mg/dL})\mu\text{mol/L}$

Collection Method Comments

Stable disease or plateau phase are no longer recommended but will be addressed by reporting time to progression (TTP) in responders.

Related Metadata References

Personal communication, IMF working group.

Blade J, Samson D, Reece D, et al. Criteria for evaluating disease response and progression in patients with multiple myeloma treated by high dose therapy and haematopoietic stem cell transplantation. Myeloma sub-committee of the EBMT. European Group for Blood and Marrow Transplant. British Journal of Haematology, 102, 1115-1123. 1998.

Multiple Myeloma – Prognostic Score (ISS)

Synonymous Names	ISS
Meteor Name	International Staging System for Multiple Myeloma
Data Element Concept	Person with cancer—primary site of cancer, code (ICD–10–AM 3rd edn) ANN{.N[N]}- haematological cancer group(Multiple Myeloma) – prognostic score (ISS)
Definition	Person with cancer – primary site of cancer - Haematological Cancer Group(Multiple myeloma)–prognostic score (ISS)
Object Class	Haematological Cancer Group
Value Domain	Prognostic score (ISS)
Guide for Use	This prognostic score uses the level of serum β 2-microglobulin and albumin to identify the level of risk at diagnosis.

Value	Stage	β 2-m (mg/L)	Risk	Median survival (Months)
1	I	< 3.5 and S Albumin \geq 35g/L	Poor	62
2	II	Neither I, III	Intermediate	44
3	III	\geq 5.5	Good	29

Collection Methods

Related Metadata

References

Greipp PR, San Miguel J, Durie BGM et al: International Staging System for Multiple Myeloma. J Clin Oncol 23:3412-3420, 2005.

Myelodysplastic Syndrome

Myelodysplastic Syndrome	
PK	Cancer Diagnosis
	Prognostic Score (IPSS) Response Date of report

Figure 9: Myelodysplastic Syndrome Structure

Myelodysplastic Syndrome – Prognostic Score (IPSS)

Synonymous Names	Prognostic Score (IPSS) Risk Score International prognostic system scoring Myelodysplastic Syndromes Risk
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Myelodysplastic Syndromes) – prognostic score (IPSS)
Data Element Concept	Person with cancer – primary site of cancer - Haematological Cancer Group(Myelodysplastic Syndrome)–prognostic score (IPSS)
Definition	The result of the prognostic score calculation for myelodysplastic syndromes for this person

	Score Value				
Prognostic Variable	0	0.5	1.0	1.5	2.0
BM Blasts (%)	<5	5-10	-	11-20	21-30
Karyotype	Good	Intermediate	Poor		
Cytopenias	0,1	2,3			

Object Class	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group(Myelodysplastic Syndromes)
Value Domain	Prognostic Score (IPSS)

Guide for Use

Parameters used: Bone marrow blast percentage, cytogenetics and cytopenias

Patients are scored (between 0 and 2) in each of the above 3 criteria depending on the severity. The total score is then used to stratify risk.

Karyotype:

Good: normal; -Y; del(5q); del(20q)

Poor: Complex (≥ 3 abnormalities), abnormal chr 7

Intermediate: all others

Cytopenias:

Defined based on number of affected lineages:

Haemoglobin $< 100\text{g/L}$

Neutrophil count $< 1.5 \times 10^9/\text{L}$

Platelet count $< 100 \times 10^9/\text{L}$

Risk

low risk: total score 0

intermediate-1 risk: total score 0.5-1.0

intermediate-2 risk: total score 1.5-2.0

high risk: total score 2.5-3.5

Outcomes:

See following table

Age related median survival by IPSS (years)					
		Low	Int-1	Int-2	High
	All Ages	5.7	3.5	1.2	0.4
Age	≤ 60	11.8	5.2	1.8	0.3
	> 60	4.8	2.7	1.1	0.5
	≤ 70	9.0	4.4	1.3	0.4
	> 70	3.9	2.4	1.2	0.4

Age related median time for 25% Evolution to AML by IPSS (years)					
		Low	Int-1	Int-2	High
	All Ages	9.4	3.3	1.1	0.2
Age	≤ 60	> 9.4 (NR)	6.9	0.7	0.2
	> 60	9.4	2.7	1.3	0.2
	≤ 70	> 9.4 (NR)	5.5	1.0	0.2
	> 70	> 5.8 (NR)	2.2	1.4	0.4

NR = not reached

Collection Methods

Comments

Related Metadata

References

Greenberg P, Cox C, LeBeau M, et al. International Scoring System for evaluating prognosis in myelodysplastic syndromes. Blood 89: 2079-88, 1997

Myelodysplastic Syndrome – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Myelodysplastic Syndrome) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Myelodysplastic Syndrome) – Response
Definition	Indicate the level of response to treatment of this patient with Myelodysplastic Syndrome
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Myelodysplastic Syndrome)
Value Domain	Response
Guide for Use	Parameters utilised: FBC and Marrow biopsy..

01 Complete Remission (CR):

- Bone marrow: $\leq 5\%$ myeloblasts with normal maturation of all cell lines*
- Persistent dysplasia will be noted*
- Peripheral blood†
 - Hgb ≥ 11 g/dL
 - Platelets $\geq 100 \times 10^9/L$
 - Neutrophils $\geq 1.0 \times 10^9/L$
 - Blasts 0%

03 Partial Remission (PR):

- All CR criteria if abnormal before treatment except:
 - Bone marrow blasts decreased by $\geq 50\%$ over pretreatment but still more than 5%.
 - Cellularity and morphology not relevant

11 Marrow Complete Remission (MaCR)

- Bone marrow: $\leq 5\%$ myeloblasts and decrease by $\geq 50\%$ over pretreatment
- Peripheral blood: if HI responses, they will be noted in addition to marrow CR

50 Stable Disease (SD):

- Failure to achieve at least PR, but no evidence of progression for > 8 weeks

61 Failure (TFI):

- Death during treatment or disease progression characterized by worsening of cytopenias, increase in percentage of bone marrow blasts, or progression to a more advanced MDS FAB subtype than pretreatment.

52 Relapse after CR or PR (RL):

- ≥ 1 of the following:
 - Return to pretreatment bone marrow blast percentage
 - Decrement of $\geq 50\%$ from maximum remission/response levels in granulocytes or platelets
 - Reduction in Hgb concentration by ≥ 15 g/L or transfusion

dependence

31 Cytogenetic Response - Complete (CCyR)

- Disappearance of the chromosomal abnormality without appearance of new ones.

34 Cytogenetic Response - Partial (PCyR):

- At least 50% reduction of the chromosomal abnormality

51 Progressive Disease (PD):

- For patients with:
 - < 5% blasts: $\geq 50\%$ increase in blasts to > 5% blasts
 - 5–10% blasts: $\geq 50\%$ increase to > 10% blasts
 - 10–20% blasts: $\geq 50\%$ increase to > 20% blasts
 - 20–30% blasts: $\geq 50\%$ increase to > 30% blasts
- Any of the following:
 - $\geq 50\%$ decrement from maximum remission/response in granulocytes or platelets
 - Reduction in Hb by ≥ 20 g/L
 - Transfusion dependence

41 Haematologic Improvement - Erythroid (HI-E):

- Pretreatment Hb < 110 g/L
- Hb increase by ≥ 15 g/d
- Relevant reduction of units of RBC transfusions by an absolute number of at least 4 RBC transfusions/ 8 weeks compared to the pretreatment transfusion number in the previous 8 weeks. Only RBC transfusions given for a Hb of 90 g/L or less pretreatment will count in the RBC transfusion response evaluation.

42 Haematologic Improvement - Platelet (HI-P):

- Pretreatment < $100 \times 10^9/L$
- Absolute increase of $\geq 30 \times 10^9/L$ for patients starting with $> 20 \times 10^9/L$ platelets
- Increase from $< 20 \times 10^9/L$ to $> 20 \times 10^9/L$ and by at least 100%

43 Haematologic Improvement - Neutrophil (HI-N):

- Pretreatment < $1.0 \times 10^9/L$
- $\geq 100\%$ increase and an absolute increase $> 0.5 \times 10^9/L$

55 Relapse after HI (HI-RL):

- ≥ 1 of the following:
 - $\geq 50\%$ decrement from maximum response levels in granulocytes or platelets
 - Reduction in Hb by ≥ 15 g/L
- Transfusion dependence

**Collection Method
Comments**

- Responses must last at least 4 weeks
- Haematologic improvement responses must last at least 8 weeks
- Pretreatment counts for haematologic improvement are averages of at least two measurements, at least 1 week apart, not influenced by transfusion.

***Related Metadata
References***

Cheson BD, Bennett JM, Kantarjian H et al. Report of an international working group to standardise response criteria for myelodysplastic syndromes. *Blood* 96, 3671-3674, 2000.

Cheson BD, Greenberg PL, Bennett JM et al. Clinical Application and Proposal for Modification of the International Working Group (IWG) Response Criteria in Myelodysplasia. *Blood First Edition Paper*, prepublished online April 11, 2006; DOI 10.1182/blood-2005-10-4149

Other Haematological Cancer

Other Haematological Cancer	
PK	<u>Cancer Diagnosis</u>
	Response
	Date of report

Figure 10: Other Haematological Cancer Structure

Other Haematological Cancer – Response

Synonymous Names	Haematological Cancer Response
Meteor Name	Person with cancer—primary site of cancer, code (ICD-10-AM 3 rd edn) ANN{.N[N]}- haematological cancer group (Other Haematological Cancer) – Response
Data Element Concept	Person with cancer—primary site of cancer, code (ICD-10-AM 3rd edn) ANN{.N[N]}- haematological cancer group (Other Haematological Cancer) – Response
Definition	Indicate the level of response to treatment of this patient with Other Haematological Cancer
Object Class	Person with cancer – primary site of cancer – haematological cancer group (Other Haematological Cancer)
Value Domain	Response
Guide for use	Select one of the values in the value domain to represent the response of this patient's other haematological cancers
Collection Methods	
Related Metadata	
References	

Appendix 1: ICD10-AM Cancer Disease Codes

C81 Hodgkin lymphoma

Includes: morphology codes M965–M966 with behaviour code /3

- C81.0 Lymphocytic predominance
Lymphocytic-histiocytic predominance
- C81.1 Nodular sclerosis
- C81.2 Mixed cellularity
- C81.3 Lymphocytic depletion
- C81.7 Other Hodgkin disease
- C81.9 Hodgkin disease, unspecified

C82 Follicular [nodular] non-Hodgkin lymphoma

Includes: follicular non-Hodgkin lymphoma with or without diffuse areas
morphology code M969 and M9591 with behaviour code /3

- C82.0 Small cleaved cell, follicular
- C82.1 Mixed small cleaved and large cell, follicular
- C82.2 Large cell, follicular
- C82.7 Other types of follicular non-Hodgkin lymphoma
- C82.9 Follicular non-Hodgkin lymphoma, unspecified
Nodular non-Hodgkin lymphoma NOS

C83 Diffuse non-Hodgkin lymphoma

Includes: morphology codes M9591, M967–M968 and M9727 with behaviour code /3

- C83.0 Small cell (diffuse)
- C83.1 Small cleaved cell (diffuse)
- C83.2 Mixed small and large cell (diffuse)
- C83.3 Large cell (diffuse)
Reticulum cell sarcoma
- C83.4 Immunoblastic (diffuse)
- C83.5 Lymphoblastic (diffuse)
- C83.6 Undifferentiated (diffuse)
- C83.7 Burkitt tumour
- C83.8 Other types of diffuse non-Hodgkin lymphoma
- C83.9 Diffuse non-Hodgkin lymphoma, unspecified

C84 Peripheral and cutaneous T-cell lymphomas

Includes: morphology code M970 and M9717–M9718 with behaviour code /3

- C84.0 Mycosis fungoides

- C84.1 Sézary disease
- C84.2 T-zone lymphoma
- C84.3 Lymphoepithelioid lymphoma
Lennert lymphoma
- C84.4 Peripheral T-cell lymphoma
- C84.5 Other and unspecified T-cell lymphomas

Note: If T-cell lineage or involvement is mentioned in conjunction with a specific lymphoma, code to the more specific description

C85 Other and unspecified types of non-Hodgkin lymphoma

Includes: morphology codes **M959** and **M967–M972** with behaviour code /3

- C85.0 Lymphosarcoma
- C85.1 B-cell lymphoma, unspecified

Note: If B-cell lineage or involvement is mentioned in conjunction with a specific lymphoma, code to the more specific description
- C85.7 Other specified types of non-Hodgkin lymphoma
Malignant:
 - Reticuloendotheliosis
 - Reticulosis
 Microglioma
- C85.9 Non-Hodgkin lymphoma, unspecified type
Lymphoma NOS
Malignant lymphoma NOS
Non-Hodgkin lymphoma NOS

C88 Malignant immunoproliferative diseases

Includes: morphology code **M976** with behaviour code /3

The following fifth character subdivision is for use with category **C88**:

- ⊗ 0 without mention of remission
- ⊗ 1 in remission

C88.0 Waldenström macroglobulinaemia

C88.1 Alpha heavy chain disease

C88.2 Gamma heavy chain disease

Franklin disease

C88.3 Immunoproliferative small intestinal disease

Mediterranean lymphoma

C88.7 Other malignant immunoproliferative diseases

C88.9 Malignant immunoproliferative disease, unspecified

Immunoproliferative disease NOS

C90 Multiple myeloma and malignant plasma cell neoplasms

Includes: morphology codes **M973** with behaviour code /3

The following fifth character subdivision is for use with category **C90**:

⊗ 0 without mention of remission

⊗ 1 in remission

C90.0 Multiple myeloma

Kahler's disease
Myelomatosis

Excludes: solitary myeloma (**C90.2**)

C90.1 Plasma cell leukaemia**C90.2 Plasmacytoma, extramedullary**

Malignant plasma cell tumour NOS
Plasmacytoma NOS
Solitary myeloma

C91 Lymphoid leukaemia

Includes: morphology codes **M982–M983, M9940** with behaviour code /3

The following fifth character subdivision is for use with category **C91**:

⊗ 0 without mention of remission

⊗ 1 in remission

C91.0 Acute lymphoblastic leukaemia

Excludes: acute exacerbation of chronic lymphocytic leukaemia (**C91.1**)

C91.1 Chronic lymphocytic leukaemia**C91.2 Subacute lymphocytic leukaemia****C91.3 Prolymphocytic leukaemia****C91.4 Hairy-cell leukaemia**

Leukaemic reticuloendotheliosis

C91.5 Adult T-cell leukaemia

C91.7 Other lymphoid leukaemia

Lymphosarcoma cell leukaemia

C91.9 Lymphoid leukaemia, unspecified**C92 Myeloid leukaemia**

Includes:leukaemia:

- granulocytic
 - myelogenous
- morphology codes **M984–M993** with behaviour code /3

The following fifth character subdivision is for use with category **C92**:

- ⊗ 0 without mention of remission
- ⊗ 1 in remission

C92.0 Acute myeloid leukaemia

Excludes: acute exacerbation of chronic myeloid leukaemia (**C92.1**)

C92.1 Chronic myeloid leukaemia**C92.2 Subacute myeloid leukaemia****C92.3 Myeloid sarcoma**

Chloroma
Granulocytic sarcoma

C92.4 Acute promyelocytic leukaemia**C92.5 Acute myelomonocytic leukaemia****C92.7 Other myeloid leukaemia****C92.9 Myeloid leukaemia, unspecified****C93 Monocytic leukaemia**

Includes:monocytoid leukaemia

morphology codes **M986** and **M989** with behaviour code /3

The following fifth character subdivision is for use with category **C93**:

- ⊗ 0 without mention of remission
- ⊗ 1 in remission

C93.0 Acute monocytic leukaemia

Excludes: acute exacerbation of chronic monocytic leukaemia (C93.1)

C93.1 Chronic monocytic leukaemia**C93.2 Subacute monocytic leukaemia****C93.7 Other monocytic leukaemia****C93.9 Monocytic leukaemia, unspecified****C94 Other leukaemias of specified cell type**

Includes: morphology codes M974, M9840, M9910 and M993–M994 with behaviour code /3

Excludes: leukaemic reticuloendotheliosis (C91.4)
plasma cell leukaemia (C90.1)

The following fifth character subdivision is for use with category C94:

⊗ 0 without mention of remission

⊗ 1 in remission

C94.0 Acute erythraemia and erythroleukaemia

Acute erythraemic myelosis
Di Guglielmo disease

C94.1 Chronic erythraemia

Heilmeyer-Schöner disease

Includes: morphology code M9950 with behaviour code /3

C94.2 Acute megakaryoblastic leukaemia

Leukaemia:
• megakaryoblastic (acute)
• megakaryocytic (acute)

C94.3 Mast cell leukaemia**C94.4 Acute panmyelosis****C94.5 Acute myelofibrosis****C94.7 Other specified leukaemias**

C95 Leukaemia of unspecified cell type

Includes: morphology code **M980** with behaviour code /3

The following fifth character subdivision is for use with category **C95**:

- ⊗ 0 without mention of remission
- ⊗ 1 in remission

C95.0 Acute leukaemia of unspecified cell type

Blast cell leukaemia
Stem cell leukaemia

Excludes: acute exacerbation of unspecified chronic leukaemia (**C95.1**)

C95.1 Chronic leukaemia of unspecified cell type**C95.2 Subacute leukaemia of unspecified cell type****C95.7 Other leukaemia of unspecified cell type****C95.9 Leukaemia, unspecified****C96 Other and unspecified malignant neoplasms of lymphoid, haematopoietic and related tissue**

Includes: morphology codes **M974–M996** with behaviour code /3

C96.0 Letterer-Siwe disease
Nonlipid:
• Reticuloendotheliosis
• Reticulosis

C96.1 Malignant histiocytosis
Histiocytic medullary reticulosis

C96.2 Malignant mast cell tumour
Malignant:
• Mastocytoma
• Mastocytosis
Mast cell sarcoma

Excludes: mast cell leukaemia (**C94.3**)
mastocytosis (cutaneous) (**Q82.2**)

C96.3 True histiocytic lymphoma

C96.7 Other specified malignant neoplasms of lymphoid, haematopoietic and related tissue

C96.9 Malignant neoplasm of lymphoid, haematopoietic and related tissue, unspecified⁷

⁷ Extracted from NCCH ICD-10-AM, July 2004, Neoplasms.

Includes: morphology code M998 with behaviour code /3

Note: Myelodysplastic syndromes have been reclassified in ICD-O Third Edition with a malignant behaviour code /3. The codes within D46 will continue to be used (although they are located in the chapter for Neoplasms of uncertain and unknown behaviour) with the appropriate morphology code as indexed.

- D46.0 Refractory anaemia without sideroblasts, so stated
- D46.1 Refractory anaemia with sideroblasts
- D46.2 Refractory anaemia with excess of blasts
- D46.3 Refractory anaemia with excess of blasts with transformation
- D46.4 Refractory anaemia, unspecified
- D46.7 Other myelodysplastic syndromes
- D46.9 Myelodysplastic syndrome, unspecified
 - Myelodysplasia NOS
 - Preleukaemia (syndrome) NOS⁸

⁸ Extracted from NCCH ICD-10-AM, July 2004, Neoplasms.

Appendix 2: Morphology Classifications

The attached is an extract from the Morphology of Neoplasms section of the International Classification of Diseases – Australian Modification – Version 10.

Morphology of Neoplasms

The third edition of the International Classification of Diseases for Oncology (ICD-O) was published in 2000. It contains a coded nomenclature for the morphology of neoplasms, which is reproduced here for those who wish to use it in conjunction with Chapter II.

The morphology code numbers consist of five digits; the first four identify the histological type of the neoplasm and the fifth, following a slash or solidus, indicates its behaviour. The one-digit behaviour code is as follows:

- /0 Benign**
- /1 Uncertain whether benign or malignant**
Borderline malignancy (*Except cystadenomas of ovary in M844–M849, which are considered to be malignant.*)
Low malignant potential (*Except cystadenomas of ovary in M844–M849, which are considered to be malignant.*)
Uncertain malignant potential
- /2 Carcinoma in-situ**
Intraepithelial
Noninfiltrating
Noninvasive
- /3 Malignant, primary site**
- /6 Malignant, metastatic site**
Malignant, secondary site
- /9 Malignant, uncertain whether primary or metastatic site**

In the nomenclature given here, the morphology code numbers include the behaviour code appropriate to the histological type of neoplasm; this behaviour code should be changed if the other reported information makes this appropriate. For example, chordoma is assumed to be malignant and is therefore assigned the code number M9370/3; the term 'benign chordoma' should, however, be coded M9370/0. Similarly, superficial spreading adenocarcinoma (M8143/3) should be coded M8143/2 when described as 'noninvasive', and melanoma (M8720/3), when described as 'secondary', should be coded M8720/6.

The following table shows the correspondence between the behaviour code and the different sections of Chapter II:

Behaviour Code	Chapter II Categories
/0 Benign neoplasms	D10–D36
/1 Neoplasms of uncertain and unknown behaviour	D37–D48
/2 In situ neoplasms	D00–D09
/3 Malignant neoplasms, stated or }	C00–C76

	presumed to be primary	}	C80–C96
/6	Malignant neoplasms, stated or presumed to be secondary		C77–C79

Occasionally a problem arises when a site given in a diagnosis is different from the site indicated by the site-specific code. In such instances, the given Chapter II code should be ignored and the appropriate code for the site included in the diagnosis should be used. For example, C50.- (breast) is added to the morphologic term Infiltrating duct carcinoma (M8500/3), because this type of carcinoma usually arises in the breast. However, if the term 'Infiltrating duct carcinoma' is used for a primary carcinoma arising in the pancreas, the correct code would be C25.9 (Pancreas, unspecified).

For neoplasms of lymphoid, haematopoietic and related tissue (M959–M998) the relevant codes from C81–C96 and D45–D47 are given. These Chapter II codes should be used irrespective of the state site of the neoplasm.

A coding difficulty sometimes arises where a morphological diagnosis contains two qualifying adjectives that have different code numbers. An example is 'transitional cell epidermoid carcinoma'. 'Transitional cell carcinoma NOS' is M8120/3 and 'epidermoid carcinoma NOS' is M8070/3. In such circumstances, the higher number (M8120/3 in this example) should be used, as it is usually more specific. For other information about the coding of morphology see the Australian Coding Standards.

CODED NOMENCLATURE FOR MORPHOLOGY OF NEOPLASMS

M800	Neoplasms, NOS
M8000/0	Neoplasm, benign
M8000/1	Neoplasm, uncertain whether benign or malignant
⊗M8000/2	Neoplasm, malignant, in situ
M8000/3	Neoplasm, malignant
M8000/6	Neoplasm, metastatic
M8000/9	Neoplasm, malignant, uncertain whether primary or metastatic
M8001/0	Tumour cells, benign
M8001/1	Tumour cells, uncertain whether benign or malignant
M8001/3	Tumour cells, malignant
⊗M8002/1	Malignant tumour, small cell type, uncertain whether benign or malignant
⊗M8002/2	Malignant tumour, small cell type, in situ
M8002/3	Malignant tumour, small cell type
⊗M8002/6	Malignant tumour, small cell type, metastatic
⊗M8002/9	Malignant tumour, small cell type, uncertain whether primary or metastatic
⊗M8003/1	Malignant tumour, giant cell type, uncertain whether benign or malignant
⊗M8003/2	Malignant tumour, giant cell type, in situ
M8003/3	Malignant tumour, giant cell type
⊗M8003/6	Malignant tumour, giant cell type, metastatic
⊗M8003/9	Malignant tumour, giant cell type, uncertain whether primary or metastatic
⊗M8004/0	Spindle cell tumour
⊗M8004/1	Malignant tumour, spindle cell type, uncertain whether benign or malignant

⊗M8004/2	Malignant tumour, spindle cell type, in situ
M8004/3	Malignant tumour, spindle cell type
⊗M8004/6	Malignant tumour, spindle cell type, metastatic
⊗M8004/9	Malignant tumour, spindle cell type, uncertain whether primary or metastatic
M8005/0	Clear cell tumour NOS
⊗M8005/1	Clear cell tumour, uncertain whether benign or malignant
⊗M8005/2	Clear cell tumour, malignant, in situ
M8005/3	Malignant tumour, clear cell type
⊗M8005/6	Malignant tumour, clear cell type, metastatic
⊗M8005/9	Malignant tumour, clear cell type, uncertain whether primary or metastatic

M959–M972 Hodgkin and Non-Hodgkin Lymphomas

<i>M959</i>	<i>Malignant lymphomas, NOS or diffuse</i>
M9590/3	Malignant lymphoma NOS
M9591/3	Lymphoma, non-Hodgkin NOS
M9596/3	Composite Hodgkin and non-Hodgkin lymphoma
<i>M965-M966</i>	<i>Hodgkin lymphoma</i>
M9650/3	Hodgkin lymphoma NOS
M9651/3	Hodgkin lymphoma, lymphocyte-rich
M9652/3	Hodgkin disease, mixed cellularity NOS
M9653/3	Hodgkin lymphoma, lymphocyte depletion NOS
M9654/3	Hodgkin lymphoma, lymphocytic depletion, diffuse fibrosis
M9655/3	Hodgkin lymphoma, lymphocytic depletion, reticular
M9659/3	Hodgkin lymphoma, nodular lymphocyte predominance
M9661/3	Hodgkin granuloma
M9662/3	Hodgkin sarcoma
M9663/3	Hodgkin lymphoma, nodular sclerosis NOS
M9664/3	Hodgkin lymphoma, nodular sclerosis, cellular phase
M9665/3	Hodgkin lymphoma, nodular sclerosis, grade 1
M9667/3	Hodgkin lymphoma, nodular sclerosis, grade 2
<i>M967-M972</i>	<i>Non-Hodgkin lymphoma</i>
<i>M967-M969</i>	<i>Mature B-cell lymphoma</i>
M9670/3	Lymphoma, small B lymphocytic NOS
M9671/3	Lymphoma, lymphoplasmacytic
M9673/3	Mantle cell lymphoma
M9675/3	Lymphoma, mixed small and large cell, diffuse
M9678/3	Primary effusion lymphoma
M9679/3	Mediastinal large B-cell lymphoma
M9680/3	Lymphoma, large B-cell, diffuse NOS
M9684/3	Lymphoma, large B-cell, diffuse, immunoblastic NOS
M9687/3	Burkitt lymphoma NOS
M9689/3	Splenic marginal zone B-cell lymphoma
M9690/3	Follicular lymphoma NOS
M9691/3	Follicular lymphoma, grade 2

M9695/3	Follicular lymphoma, grade 1
M9698/3	Follicular lymphoma, grade 3
M9699/3	Marginal zone B-cell lymphoma NOS
<i>M970-M971</i>	<i>Mature T- and NK-cell lymphomas</i>
M9700/3	Mycosis fungoides
M9701/3	Sezary syndrome
M9702/3	Mature T-cell lymphoma NOS
M9705/3	Angioimmunoblastic T-cell lymphoma
M9708/3	Subcutaneous panniculitis-like T-cell lymphoma
M9709/3	Cutaneous T-cell lymphoma NOS
M9714/3	Anaplastic large cell lymphoma, T cell and Null cell type
M9716/3	Hepatosplenic gamma-delta cell lymphoma
M9717/3	Intestinal T-cell lymphoma
M9718/3	Primary cutaneous CD30+ T-cell lymphoproliferative disorder
M9719/3	NK/T-cell lymphoma, nasal and nasal-type
<i>M972</i>	<i>Precursor cell lymphoblastic lymphoma</i>
M9727/3	Precursor cell lymphoblastic lymphoma NOS
M9728/3	Precursor B-cell lymphoblastic lymphoma
M9729/3	Precursor T-cell lymphoblastic lymphoma
M973	Plasma Cell Tumours
M9731/3	Plasmacytoma NOS
M9732/3	Multiple myeloma
M9733/3	Plasma cell leukaemia
M9734/3	Plasmacytoma, extramedullary
M974	Mast Cell Tumours
M9740/1	Mastocytoma NOS
M9740/3	Mast cell sarcoma
M9741/3	Malignant mastocytosis
M9742/3	Mast cell leukaemia
M975	Neoplasms of Histiocytes and Accessory Lymphoid Cells
M9750/3	Malignant histiocytosis
M9751/1	Langerhans cell histiocytosis NOS
M9752/1	Langerhans cell histiocytosis, unifocal
M9753/1	Langerhans cell histiocytosis, multifocal
M9754/3	Langerhans cell histiocytosis, disseminated
M9755/3	Histiocytic sarcoma
M9756/3	Langerhans cell sarcoma
M9757/3	Interdigitating dendritic cell sarcoma
M9758/3	Follicular dendritic cell sarcoma
M976	Immunoproliferative Diseases
M9760/3	Immunoproliferative disease NOS
M9761/3	Waldenstrom macroglobulinaemia
M9762/3	Heavy chain disease NOS
M9764/3	Immunoproliferative small intestinal disease

M9765/1	Monoclonal gammopathy of undetermined significance
M9766/1	Angiocentric immunoproliferative lesion
M9767/1	Angioimmunoblastic lymphadenopathy
M9768/1	T-gamma lymphoproliferative disease
M9769/1	Immunoglobulin deposition disease

M980–M994 Leukaemias

<i>M980</i>	<i>Leukaemias, NOS</i>
M9800/3	Leukaemia NOS
M9801/3	Acute leukaemia NOS
M9805/3	Acute biphenotypic leukaemia
<i>M982-M983</i>	<i>Lymphoid leukaemias</i>
M9820/3	Lymphoid leukaemia NOS
M9823/3	B-cell chronic lymphocytic leukaemia/small lymphocytic lymphoma
M9826/3	Burkitt cell leukaemia
M9827/3	Adult T-cell leukaemia/lymphoma
M9831/3	T-cell large granular lymphocytic leukaemia
M9832/3	Prolymphocytic leukaemia NOS
M9833/3	Prolymphocytic leukaemia, B-cell type
M9834/3	Prolymphocytic leukaemia, T-cell type
M9835/3	Precursor cell lymphoblastic leukaemia NOS
M9836/3	Precursor B-cell lymphoblastic leukaemia
M9837/3	Precursor T-cell lymphoblastic leukaemia
<i>M984-M993</i>	<i>Myeloid leukaemias</i>
M9840/3	Acute myeloid leukaemia, M6 type
M9860/3	Myeloid leukaemia NOS
M9861/3	Acute myeloid leukaemia NOS
M9863/3	Chronic myeloid leukaemia NOS
M9866/3	Acute promyelocytic leukaemia
M9867/3	Acute myelomonocytic leukaemia
M9870/3	Acute basophilic leukaemia
M9871/3	Acute myeloid leukaemia with abnormal marrow eosinophils
M9872/3	Acute myeloid leukaemia, minimal differentiation
M9873/3	Acute myeloid leukaemia without maturation
M9874/3	Acute myeloid leukaemia with maturation
M9875/3	Chronic myelogenous leukaemia, BCR/ABL positive
M9876/3	Atypical chronic myeloid leukaemia, BCR/ABL negative
M9891/3	Acute monocytic leukaemia
M9895/3	Acute myeloid leukaemia with multilineage dysplasia
M9896/3	Acute myeloid leukaemia, t(8;21)(q22;q22)
M9897/3	Acute myeloid leukaemia, 11q23 abnormalities
M9910/3	Acute megakaryoblastic leukaemia
M9920/3	Therapy-related acute myeloid leukaemia NOS
M9930/3	Myeloid sarcoma
M9931/3	Acute panmyelosis with myelofibrosis

<i>M994</i>	<i>Other leukaemias</i>
M9940/3	Hairy cell leukaemia
M9945/3	Chronic myelomonocytic leukaemia NOS
M9946/3	Juvenile myelomonocytic leukaemia
M9948/3	Aggressive NK-cell leukaemia
M995–M996	Chronic Myeloproliferative Disorders
M9950/3	Polycythaemia vera
M9960/1	Chronic myeloproliferative disease
M9960/3	Chronic myeloproliferative disease NOS
M9961/3	Myelosclerosis with myeloid metaplasia
M9962/3	Essential thrombocythaemia
M9963/3	Chronic neutrophilic leukaemia
M9964/3	Hypereosinophilic syndrome
M997	Other Haematologic Disorders
M9970/1	Lymphoproliferative disorder NOS
M9975/1	Myeloproliferative disease NOS
M998	Myelodysplastic Syndromes
M9980/3	Refractory anaemia
M9982/3	Refractory anaemia with sideroblasts
M9983/3	Refractory anaemia with excess of blasts
M9984/3	Refractory anaemia with excess blasts in transformation
M9985/3	Refractory cytopenia with multilineage dysplasia
M9986/3	Myelodysplastic syndrome with 5q deletion syndrome
M9987/3	Therapy-related myelodysplastic syndrome NOS
M9989/3	Myelodysplastic syndrome NOS ⁹

⁹ Extracted from NCCH ICD-10-AM, July 2004, Appendix A: Morphology of Neoplasms.