

National Disease Registration Service (NDRS)

Key Performance Indicators:
Haematology Staging 101
v1 June 2024

Welcome to this NDRS training module, which has been designed as a quick guide to Haematology Staging

Haematology Staging 101: What is Stage?

Haematology stage is an assessment of how severe or advanced the disease is. A patient can and will be staged at many points within their pathway from diagnosis, pre-treatment, post-treatment and in follow-up

It's important to assess the stage of a cancer as it tells the clinical team how severe the disease is. A patient may be staged at many points in their pathway.

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The Principles of using a staging system

Enables the practice of classifying cancer cases into groups according to their anatomical extent at the point of diagnosis. It is extremely important as it;

- Helps the clinician in the **planning of the treatment**
- Gives an indication of the **prognosis for survival** for primary disease
- Assists in the evaluation of the results of the treatment and outcome analysis
- Supports cancer-controlled activities
- Helps with the development and implementation of **clinical guidelines and policy**

Clinically, a staging system is used to plan the patient's care, to indicate the likely prognosis, and to evaluate the effectiveness of certain treatments. Knowing the stage is also important when developing and implementing clinical guidelines.

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For COSD, haematology stage is collected only for defined haematological malignancies which are new primary diagnoses, at the point of diagnosis

However, for the purposes of COSD, Haematology stage is only collected for specified haematological malignancies that are new primary diagnoses and only at the point of diagnosis

Haematology Staging 101: Staging Systems

Haematology Cancers

Hodgkin's Lymphoma	Ann Arbor Stage
Non-Hodgkin's Lymphoma (Adult)	Ann Arbor Stage
Non-Hodgkin's Lymphoma (CTYA)	Murphy / St Jude's Staging System
Myeloma	Revised International Staging System (R-ISS)
Chronic Lymphocytic Leukaemia	BINET Stage

Other Haematological diseases are not considered stageable

While Haematology staging is usually referred to as being “site-specific”, it’s actually **disease-specific**, and in one case, age-specific! For instance, all lymphomas in adults and Hodgkin’s lymphomas in CTYA patients are staged using the Ann Arbor system. But **non-Hodgkin’s** lymphoma in CTYA patients is staged using the Murphy / St Jude’s stage. Myeloma requires an R-ISS stage and Chronic Lymphocytic Leukaemia needs a Binet stage. Other Haem diseases are not considered to be stageable.

Lymphoma – Stage – Hodgkin Lymphoma in children and all adult lymphomas

The Ann Arbor Staging Classification is used for all Lymphomas in adults (and for Hodgkin Lymphomas in children)

I	One region of lymph nodes, or spleen or thymus or Waldeyer's ring enlarged
II	Two regions of lymph nodes enlarged on the same side of diaphragm
III	Lymph nodes enlarged on both sides of diaphragm
IV	Disease outside lymph nodes e.g. liver, bone marrow

As I mentioned, all Lymphomas in adults and Hodgkin Lymphomas in children are staged using the Ann Arbor staging system, much of which revolves around the number and placement of enlarged lymph nodes ...

Ann Arbor Symptoms

Additional stage designation based on presence / absence of specific symptoms

A

No Symptoms

B

Presence of any of the following: unexplained persistent or recurrent fever (greater than 38°C / 101.5°F), drenching night sweats, unexplained weight loss of 10% or more within the last 6 months)

If the patient is displaying a raised temperature, severe night sweats or a significant unexplained weight loss, this would need to be noted as Answer B under Ann Arbor symptoms

Ann Arbor Extranodality

Additional staging designation based on extranodal involvement. Code "E" if there is involvement of a single extranodal (other than the lymph nodes) site that directly adjoins or is next to the known nodal group

E

Extranodal involvement

O

No Extranodal involvement

If lymphoma is found outside the nodes this would be recorded as Answer E under Extranodality

Ann Arbor Bulk

Additional staging designation based on presence of bulky disease.

Code "X" if there is presence of "bulky" disease, that is, a nodal mass whose greatest dimension is more than 10 centimetres in size, and/or a widening of the mediastinum (middle chest) by more than one-third.

X

Yes, "bulky" disease present

0

No "bulky" disease present

Where Bulky disease is found, record Answer X under Ann Arbor Bulk

Ann Arbor Splenic Involvement

Additional staging designation based on splenomegaly or normal spleen size with confirmed disease involvement. Code 'S' if either is true.

S	Spleen involvement or splenomegaly
0	No Spleen involvement or splenomegaly

If an enlarged Spleen (or Splenomegaly) is found, record Answer S under Splenic Involvement

Lymphoma – Stage – Non-Hodgkin Lymphoma in CTYA

MURPHY ST. JUDE STAGING SYSTEM

STAGE	Criteria for Extent of Disease
I	A single tumour (extranodal) or single anatomic area (nodal) with the exclusion of mediastinum or abdomen.
II	A single tumour (extranodal) with regional node involvement. Two or more nodal areas on the same side of the diaphragm. Two single (extranodal) tumours with or without regional node involvement on the same side of the diaphragm. A primary gastrointestinal tract tumour, usually in the ileocecal area, with or without involvement of associated mesenteric nodes only, grossly completely resected.
III	Two single tumours (extranodal) on opposite sides of the diaphragm. Two or more nodal areas above and below the diaphragm. All the primary intrathoracic tumours (mediastinal, pleural, thymic). All extensive primary intraabdominal disease, unresectable. All paraspinal or epidural tumours, regardless of other tumour site(s).
IV	Any of the above with initial CNS and/or bone marrow involvement.

The Murphy St Jude system is used for Non-Hodgkin Lymphoma in CTYA patients

Myeloma - Stage

- Myeloma is staged using the Revised International Staging System (**R-ISS**) for Myeloma.

Stage 1

Beta-2 microglobulin ≤ 3.5 g/dL and albumin ≥ 3.5 g/dL. Standard risk CA by iFISH. Normal LDH

Stage 2

Does not meet the criteria for Stage 1 or Stage 3.

Stage 3

Beta-2 microglobulin of ≥ 5.5 g/dL, **and either** High risk CA by iFISH **or** high LDH

Myeloma requires an R-ISS stage. This is based on lab results and molecular testing.

Leukaemia - Stage

Most types of leukaemia are not stageable

Only Chronic Lymphocytic Leukaemia (CLL) is staged using the Binet system

Binet stage is derived from multiple diagnostic test results:

- Platelet count
- Hb (haemoglobin count)
- Lymphadenopathy (enlarged lymph nodes)
- Hepatomegaly (enlarged liver)
- Splenomegaly (enlarged spleen)

Most leukaemias are not stageable. The only type of leukaemia that requires a stage is Chronic Lymphocytic Leukaemia, or CLL.

Leukaemia – Stage - Binet

The Binet stage is supplied by the clinical team

Stage A

If Platelet count $> 99 \times 10^9/L$ and Hb >99 and 0, 1 or 2 areas of organ enlargement (number of lymph node groups plus score 1 for hepatomegaly, 1 for splenomegaly)

Stage B

If Platelet count > 99 and Hb >99 and 3, 4 or 5 areas of organ enlargement

Stage C

If Hb <100 or platelet count <100

Binet stage for CLL relies on blood tests, lymphadenopathy and any enlargement of the liver or spleen

COSD Haematological Stage Completeness

A stageable haematological cancer is considered staged when the site-specific stage has been included in the COSD submission

All stage **MUST** have the following data items completed for the stage to be included in the COSD submission extract:

- Reported Date
- Reporting Organisation

Please remember to include both the reporting date and the Reporting organisation. Without these data items, the haematology stage will not be included in the COSD submission.

The Data Liaison Team

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And of course, we're here to support you. If you have any questions about Haematology staging or any other aspect of COSD data, do please get in touch with your regional data liaison manager