

National Disease Registration Service (NDRS)

Haematology – Leukaemia
v4 August 2024



Welcome to this NDRS training module on Haematology – Leukaemia, which has been designed to help Cancer Administration staff gain a better understanding of Leukaemia, the terminology used by the clinical teams and where to find guidance on the correct codes.

Agenda

- Introduction
- Leukaemia
- Summary
- Acknowledgements

This module may be paused at any time

It is advised that this module is viewed in conjunction with Haematology – An Introduction, and the searchable PDF: Haematology Appendix A, Classification



We're going to give you a brief introduction to Leukaemia including some of the symptoms that patients might experience. We'll then look at the diagnosis & treatment options. This module can be paused at any time. A PDF of these slides is also available for reference.

Leukaemia

In this section we will cover:

- Types of Leukaemia
- Causes & Risk Factors
- Signs & Symptoms
- Diagnosis
- Morphology
- ICD10 Classification
- Stage
- Treatment



Firstly, we'll look at some of the types of leukaemia...

Leukaemia – Types of Leukaemia

- Leukaemia occurs in several forms and these are often referred to using an acronym. These may include:
 - AML – Acute Myeloid Leukaemia
 - ALL – Acute Lymphoblastic Leukaemia
 - CML – Chronic Myeloid Leukaemia
 - CLL – Chronic Lymphocytic Leukaemia



Leukaemia occurs in a number of forms, many of which are referred to using acronyms including AML, ALL, CML and CLL.

Leukaemia – Causes & Risk Factors

- Radiation exposure
- Smoking
- Chemical exposure
- Genetics – some inherited conditions increase the risk of leukaemia
- Blood disorders – including myelodysplastic syndrome or myeloproliferative disorders
- Autoimmune conditions – including rheumatoid arthritis and ulcerative colitis
- Age – depending on the type of leukaemia it may be more common in older or younger people



Whilst the cause of most leukaemias is not fully known, some risk factors have been identified. These include radiation exposure, smoking, autoimmune conditions and age

Leukaemia – Signs & Symptoms

Some signs and symptoms apply to all types of leukaemia:

- Fatigue
- Pain
- Breathlessness on exertion



Leukaemias often present with fatigue, pain and breathlessness...

Leukaemia – Signs & Symptoms

Symptoms specific to a particular type of malignancy include:

	AML	ALL	CML	CLL
Frequent Infections	X	X		X
Enlarged Lymph Nodes		X		X
Bruising and Bleeding	X	X		
Sweats			X	X
Weight Loss			X	
Abdominal Discomfort			X	



... while some symptoms are specific to the type of leukaemia involved.

Leukaemia – Diagnosis

The diagnosis of leukaemia usually requires a combination of multiple tests. These may include:

- Full blood count (FBC)
- Bone marrow aspirate and bone marrow trephine biopsy
- Immunophenotype on marrow or blood cells (a process that uses antibodies to identify cells based on the types of antigens or markers on the surface of the cells)
- Cytogenetics on marrow or blood (the examination of the structure and function of chromosomes)
- Molecular markers on marrow or blood
- Lumbar puncture (for ALL)
- Ultrasound (sometimes for CML and CLL)

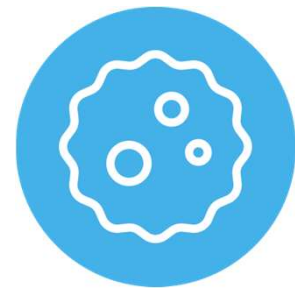


In order to reach a diagnosis of Leukaemia, a range of tests is usually needed. These might include a full blood count, bone marrow aspirate or a bone marrow trephine. Sometimes, other tests are required such as immunophenotyping

Leukaemia – Morphology

In acute leukaemia, the abnormalities are due to immature blood cells or blasts - cells which are not able to function normally

- There is a rapid growth and progression of these useless cells preventing the formation of healthy blood cells and, without treatment, the patient may die within days or a few weeks



In chronic leukaemia the blood cells will develop into mature cells but are abnormal in form and unable to function well



Leukaemias are sometimes classified as Acute, where the abnormalities in the blood cells are very severe. Untreated, the patient may die in a short space of time. In Chronic leukaemia, the cellular abnormalities are less pronounced, meaning the blood cells can retain partial functionality

Leukaemia – Morphology

The ICD-O-3 morphology code may vary depending on the specific diagnostic test results

- Always refer to the clinical team for guidance on the exact diagnosis.

Please refer to the searchable reference list provided in: NDRS PDF: Haematology – Appendix A, Classifications for the relevant ICD-O-3 morphology code to use

Appendix A - Classification

ICD10_DATALIST	ICD10_CODE	ICD10_TERM_DESC	ICD10_CODE	ICD10_DESC
ALL	9812/3 A	B-lymphoblastic leukaemia with t(9;22)(q34;q11.21) BCR-ABL1	C83.5	Lymphoblastic (diffuse) lymphoma
ALL	9811/3 A	B-lymphoblastic lymphoma with t(11;14)(p23;q24) MLL	C83.5	Lymphoblastic (diffuse) lymphoma
ALL	9811/3 A	B-lymphoblastic lymphoma, NOS	C83.5	Lymphoblastic (diffuse) lymphoma
DLBCL	9880/3 C	Burkitt lymphoma	C83.3	Diffuse large B-cell lymphoma
Other lymphomas	9566/3	B-cell lymphoma, intermediate between DLBCL and follicular nodules	C85.1	B-cell lymphoma, unspecified
CD34 ONLY	9823/3	B-cell prolymphocytic leukaemia	C81.3	Prolymphocytic leukaemia of B-cell type
Other lymphomas	9522/3	Blastic plasmacytoid dendritic cell neoplasm	C86.4	Blastic NK-cell lymphoma
ALL	9826/3	Burkitt cell leukaemia	C81.8	Mature B-cell leukaemia Burkitt type
Other lymphomas	9807/3	Burkitt lymphoma	C83.7	Burkitt lymphoma
CD34 ONLY	9864/3	Chronic eosinophilic leukaemia, NOS	C87.5	Chronic eosinophilic leukaemia (hypereosinophilic syndrome)
CEL	9823/3	Chronic lymphocytic leukaemia/small lymphocytic lymphoma	C81.1	Chronic lymphocytic leukaemia of B-cell type

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Leukaemias have specific morphology codes. Your clinical team will provide an exact diagnosis description which will enable you to determine the relevant codes from the searchable PDF reference list contained in Appendix A, Classification. Instructions on downloading this are included in the Summary.

Leukaemia – ICD10 Classification

There are many types of leukaemia but the main types are grouped by the white blood cell type from which it arises i.e. myeloid cells or lymphoid cells
Within each category, the behaviour of the leukaemia is subdivided into acute or chronic. Broadly, the diseases are classified in their main groups :

Myeloid

- Acute myeloid leukaemia
- Chronic myeloid leukaemia

Lymphoid

- Acute lymphoblastic leukaemia
- Chronic lymphocytic leukaemia



While there are many subtypes of leukaemia, broadly speaking they are classified according to cell type and severity. However...

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)



Leukaemia is not generally stageable, the exception being CLL which is staged. Binet staging for CLL relies on a number of factors, including platelet count and the presence of an enlarged liver or spleen

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

For more details, please see the latest COSD User Guide ([link in the summary](#)). Depending on your cancer data management system, you may be able to enter the stage grouping directly or it may be necessary to enter the individual test results to enable the system to calculate the stage



Depending on your cancer management system, you may be able to enter the stage as supplied by your clinical team or it may be necessary to enter individual parameters to enable the system to calculate the stage

Leukaemia - Stage

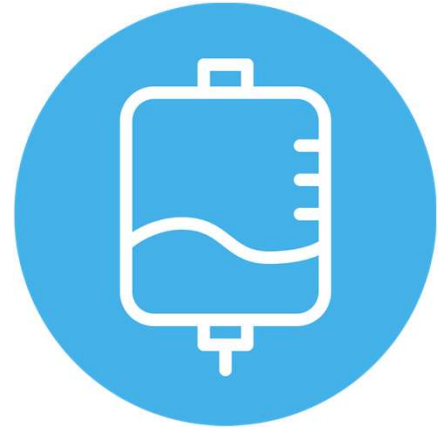
- All Site-specific stage **MUST** have the below data items completed to enable to the stage to be reported to the Registry
- Reported Date
- Reporting Organisation



It's important that the reported date and reporting organisation are also recorded to ensure that the site specific stage is included in the COSD submission

Leukaemia –Treatment- Chemotherapy

- Drug therapy is the main form of treatment for haematological disease
- By combining drugs which act in different ways against the malignant cells, the effectiveness of the chemotherapy can be improved and may reduce the risk of chemotherapy resistance to a single drug

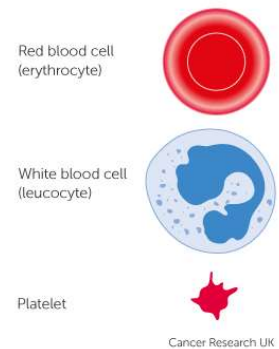


Chemotherapy is the usual treatment for leukaemia, often using different drugs in combination

Leukaemia –Treatment – Stem Cell Transplants

Stem cells are normally found in the bone marrow with small numbers found in the blood

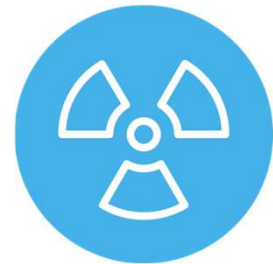
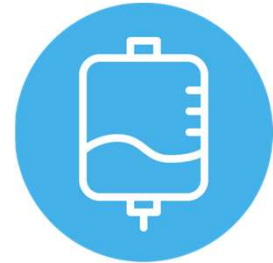
- Bone marrow transplants (BMT) and peripheral blood stem cell transplants (PBSCT) are intensive treatments that may be used to treat people with leukaemia, myeloma or lymphoma
- The purpose of a stem cell transplant is to replace abnormal cells with healthy cells developed from the donor stem cells



Some patients may be offered a stem cell transplant, the purpose of which is to replace the abnormal cells with healthy versions that will develop from the new stem cells

Leukaemia –Treatment – Stem Cell Transplants

- Chemotherapy and/or radiotherapy are used prior to the transplant with the aim of killing malignant cells and (if donor cells are being used in the transplant) suppress the patient's immune system, preventing rejection of the transplant
- Stem cell transplants can be:
 - Autologous transplant (using the patient's own stem cells)
 - Allogeneic transplant (using donor stem cells)



Prior to the transplant, chemotherapy, sometimes in combination with radiotherapy, will be used to destroy the patient's malignant cells. Where donor cells are used, the patient's immune system may need to be suppressed to prevent rejection.

Summary



In summary...

Summary

- There are many sub-types of leukaemia but the most common types are categorised as AML, CML, ALL or CLL



Leukaemias are broadly categorised as AML, CML, ALL or CLL

Summary

- There are many sub-types of leukaemia but the most common types are categorised as AML, CML, ALL or CLL
- Diagnosing leukaemia relies on a wide range of tests which may include blood tests, bone marrow biopsies or imaging



Diagnostic tests for leukaemia may include blood tests and bone marrow biopsies

Summary

- There are many sub-types of leukaemia but the most common types are categorised as AML, CML, ALL or CLL
- Diagnosing leukaemia relies on a wide range of tests which may include blood tests, bone marrow biopsies or imaging
- Once diagnosed, most forms of leukaemia are not stageable with the exception of CLL which uses the Binet staging system - please ensure that the staging date and organisation are also recorded



Only CLL requires a stage to be recorded. Other types of leukaemia are not considered stageable

Summary

- There are many sub-types of leukaemia but the most common types are categorised as AML, CML, ALL or CLL
- Diagnosing leukaemia relies on a wide range of tests which may include blood tests, bone marrow biopsies or imaging
- Once diagnosed, most forms of leukaemia are not stageable with the exception of CLL which uses the Binet staging system - please ensure that the staging date and organisation are also recorded
- Treatment for leukaemia is usually chemotherapy but sometimes a stem cell transplant is offered. This would require chemotherapy and/or radiotherapy prior to the transplant



Chemotherapy is the usual treatment for leukaemia although stem cell transplants may be offered

Summary

- The classification of Haematological disease is complex and relies on multiple factors. Your clinical team will be able to provide you with the exact diagnosis description. The Haematology Appendix A, Classification will guide you on the codes to record for ICD10 and ICD-O-3 morphology



Haematology classification is complicated and usually relies on a number of test results to allow the clinical team to provide you with an exact diagnosis description. The searchable PDF: Haematology Appendix A, Classification will guide you to the correct ICD10 diagnosis- and ICD-O-3 morphology-codes for that diagnosis.

Summary

- The classification of Haematological disease is complex and relies on multiple factors. Your clinical team will be able to provide you with the exact diagnosis description. The Haematology Appendix A, Classification will guide you on the codes to record for ICD10 and ICD-O-3 morphology
- If a Haematological disease is diagnosed it may or may not be classified as malignant. While all haematological malignancies must be recorded, please refer to the COSD User Guide (Haematology section) for the list of D or E coded conditions that must also be recorded



Not all haematological conditions are C coded as malignant in ICD10. While all C coded haematological disease must be recorded, please refer to the Haematology section of the COSD user guide for the other conditions that require a COSD record.

Summary

- Additional guidance on recording COSD data including morphology, topography, staging and recording a diagnosis can be found at: <https://digital.nhs.uk/ndrs/data/cancer-data-training-materials>
- For Haematology, this includes an Introduction plus disease specific modules for Lymphoma and Myeloma as well as a searchable PDF: Haematology - Appendix A, Classification
- Staging data sheets can also be downloaded from the NDRS website for clinical use: <https://digital.nhs.uk/ndrs/data/cancer-data-training-materials/staging-sheets>

Additional training modules as well as Staging sheets for clinical use may be downloaded from the NDRS website.

Summary

- If in any doubt as to whether you should be recording a haematology diagnosis, please refer to the latest COSD User Guide, Appendix C
- For guidance on the required staging system, please refer to the latest COSD User Guide, Appendix E
- <https://digital.nhs.uk/ndrs/data/data-sets/cosd#downloads>



Do please remember, guidance **is** available on our website. You can download the COSD User Guide by clicking on this link and selecting the COSD version appropriate to your trust.

Acknowledgements

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If you have any questions on the information contained within this module or about COSD in general, do please feel free to email your regional Data Liaison Manager