

National Disease Registration Service (NDRS)

Haematology – Myeloma
v4 August 2024



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

Agenda

- Introduction
- Myeloma
- 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 Myeloma 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.

Myeloma

In this section we will cover:

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



We'll start off by looking at Causes & Risk factors...

Myeloma – Causes & Risk Factors

- Age – more common in people over 40
- Gender – slightly more common in men than in women
- Obesity – overweight and obese people have a higher risk of myeloma
- MGUS - Monoclonal Gammopathy of Undetermined Significance (a rare precursor condition relating to a protein in the blood)
- Family history
- Impaired immune system



... which for plasma cell neoplasms such as Myeloma include Age, body weight and an impaired immune system

Myeloma – Signs & Symptoms

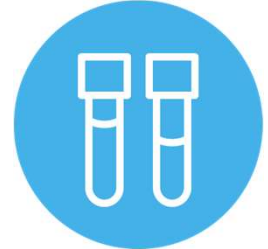
- Anaemia (reduced red blood cell count)
- Neutropenia (reduced neutrophil count, neutrophils are a type of white blood cell and form part of the immune system)
- Thrombocytopenia (low platelet count reducing the blood's ability to form clots in order to seal wounds)
- Bone pain
- Fractures (may be due to reduced calcium levels in the bones)
- Hypercalcaemia (high calcium levels in the blood)
- Kidney failure
- Repeated infections, especially chest infections



Myeloma often presents with symptoms including anaemia, bone fractures and repeated infections, especially chest infections

Myeloma - Diagnosis

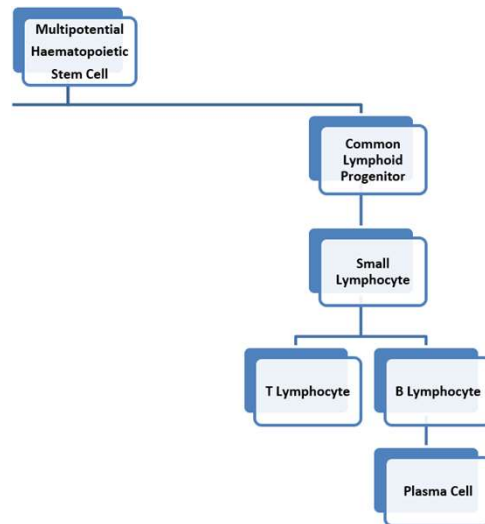
- Full blood count
- Skeletal survey
- Bone marrow sample
- Blood Immunoglobulin levels (a group of proteins in the blood)
- Paraprotein measurement (a particular type of immunoglobulin)
- Serum calcium (serum forms part of the blood)
- Creatinine (to determine kidney function)
- Albumin (a protein made by the liver)
- Beta-2 microglobulin (a small protein normally found on the surface of cells including some white blood cells, which is sometimes found in blood or urine)
- Urine and blood light chain levels (part of the antibodies, an element of the immune system)



The diagnosis of myeloma often requires a number of blood and urine tests which might look at cell counts or the levels of various proteins, as well as radiological imaging of the skeleton.

Myeloma – ICD10 Classification & Morphology

- Myeloma and other plasma cell neoplasm develop from plasma cells which are descended from B lymphocytes
- Plasma cells secrete Immunoglobulins (certain types of proteins) or antibodies which circulate in the blood and can be detected in a blood sample to help determine the specific type of neoplasm



Myeloma is a form of plasma cell neoplasm, meaning it's derived from the lymphoid cell line. Certain types of protein are secreted by plasma cells which can be detected by a blood test to help determine the specific neoplasm type.

Myeloma – ICD10 Classification & Morphology

Types of Plasma Cell Neoplasm

- Myeloma affects multiple places in the body where bone marrow is normally active in an adult i.e. within the bones of the spine, skull, pelvis, ribs and the areas around the shoulders and hips
- Solitary plasmacytoma of bone SPB (a single malignant plasma cell tumour occurring in the bone)
- Extramedullary plasmacytoma EMP (arises in tissue other than bone, most EMPs are in the head and neck area)



Myeloma affects multiple areas of active bone marrow in the body ... whereas solitary plasmacytomas occur singly. Extramedullary plasmacytomas arise outside the skeletal structure

Myeloma – ICD10 Classification & Morphology

- The ICD10 code and ICD-O-3 morphology will vary depending on the specific diagnostic test results – always refer to the clinical team for guidance on the exact diagnosis description
- Please refer to the searchable reference list provided in the searchable PDF: Haematology – Appendix A, Classifications for the relevant ICD-O-3 morphology & ICD10 codes to use

Appendix A - Classification

ICD10_CATEGORY	ICD10_CODE	ICD10_DESCRIPTION	ICD10_CODE	ICD10_DESCRIPTION
ALL	9812/7 A	B lymphoblastic lymphoma with 10-22% CD44 (L11.2) (ICD-O-3)	C83.5	Lymphoblastic (diffuse) lymphoma
ALL	9813/7 A	B lymphoblastic lymphoma with 10-22% CD44 (L11.2) (ICD-O-3)	C83.5	Lymphoblastic (diffuse) lymphoma
ALL	9813/7 B	B lymphoblastic lymphoma, NOS	C83.5	Lymphoblastic (diffuse) lymphoma
DLBCL	9860/3 C	B-cell lymphoma, intermediate between DLBCL and BL	C83.3	Diffuse large B-cell lymphoma
DLBCL	9860/3 C	B-cell lymphoma, intermediate between DLBCL and BL	C83.3	Diffuse large B-cell lymphoma
Other Lymphoma	9796/7	DLBCL, CD44 of Hodgkin	C85.1	B-cell lymphoma, unspecified
CDCL ONLY	9813/2	B-cell and lymphocytic leukaemia	C91.1	Prolymphocytic leukaemia of B-cell type
Other Lymphoma	9727/2	Blastic plasmacytoid dendritic cell neoplasm	C86.4	Blastic PLD-cell lymphoma
ALL	9856/2	Burkitt's leukaemia	C81.4	Monoclonal B-cell leukaemia Burkitt type
Other Lymphoma	9857/2	Burkitt's lymphoma	C83.7	Burkitt lymphoma
CDCL ONLY	9864/2	Chronic eosinophilic leukaemia, NOS	D47.5	Chronic eosinophilic leukaemia (per eosinophilic leukaemia)
CLL	9823/7	Chronic lymphocytic leukaemia/small lymphocytic lymphoma	C91.3	Chronic lymphocytic leukaemia of B-cell type

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Both the ICD10 code and the ICD-O-3 morphology code may vary depending on the particular type of neoplasm. 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, Classifications. Instructions on downloading this are included in the Summary.

Myeloma - Stage

- Myeloma is staged using the International Staging System (ISS) or the Revised International Staging System (R-ISS) for Myeloma. **R-ISS** is currently recordable for COSD. ISS is derived from Beta2 Microglobulin and Albumin lab results. R-ISS uses additional data (Chromosomal Abnormalities & Lactate Dehydrogenase)

ISS

Stage 1

Beta 2 M less than 3.5 and Albumin greater than 34

Stage 2

Beta 2 M less than 3.5 **and** albumin less than 35, **OR** Beta 2 M 3.5 - 5.5

Stage 3

Beta 2 M greater than 5.5

R-ISS

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

The Revised International Staging System ... or R-ISS ... is recordable for COSD versions 9 & 10.

Myeloma - 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

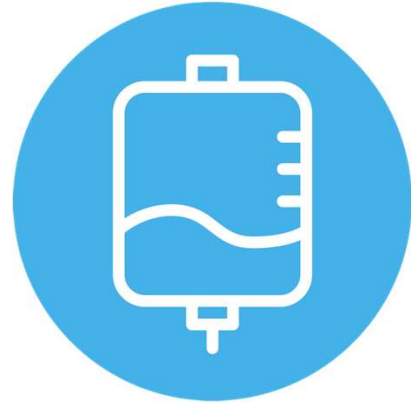
Myeloma – Smouldering myeloma

- Where a diagnosis of smouldering / asymptomatic myeloma is made, this may be based on raised serum or urinary monoclonal protein levels and therefore include patients who do not have a positive bone marrow
- As the R-ISS stage includes a FISH assessment for chromosomal abnormalities performed on the bone marrow, it may not be possible to assign an R-ISS stage to some patients with smouldering or asymptomatic myeloma
- Where smouldering / asymptomatic myeloma is diagnosed there is currently no specific morphology code for it in ICD-O-3. To differentiate this form of myeloma, please note the diagnosis as smouldering / asymptomatic in the Primary Diagnosis Subsidiary Comment field in your cancer data management system

A smouldering / asymptomatic myeloma diagnosis may be made where the patient does not have bone marrow involvement. This means it may not be possible to stage the disease using R-ISS. Where smouldering / asymptomatic myeloma is diagnosed, please note this in the Primary Diagnosis Subsidiary Comment field in your cancer data management system.

Myeloma – 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

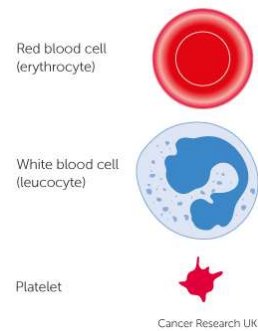


Chemotherapy is a common treatment for myeloma, often using different drugs in combination

Myeloma – Treatment – Stem Cell Transplants

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

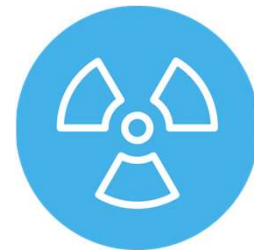
- Bone marrow transplants (BMT) and peripheral blood stem cell transplants (PBSCT) are intensive treatments that may be used to treat people with leukaemia, myeloma or lymphoma
- The purpose of a stem cell transplant is to replace abnormal cells with healthy cells developed from the donor stem cells



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

Myeloma – 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 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 over several different types of plasma cell neoplasms, one of which is myeloma



Myeloma is a form of plasma cell neoplasm

Summary

- There are over several different types of plasma cell neoplasms, one of which is myeloma
- Diagnosing myeloma relies on a wide range of tests which may include blood tests, urine tests and imaging



The diagnostic pathway may include multiple blood and urine tests as well as imaging

Summary

- There are over several different types of plasma cell neoplasms, one of which is myeloma
- Diagnosing myeloma relies on a wide range of tests which may include blood tests, urine tests and imaging
- Myeloma is staged for COSD version 9 using the R-ISS system – please ensure that the staging date and organisation are also recorded



COSD version 9 requires the use of R-ISS staging for myeloma

Summary

- There are over several different types of plasma cell neoplasms, one of which is myeloma
- Diagnosing myeloma relies on a wide range of tests which may include blood tests, urine tests and imaging
- Myeloma is staged for COSD version 9 using the R-ISS system – please ensure that the staging date and organisation are also recorded
- Treatment for myeloma is usually chemotherapy but sometimes a stem cell transplant is offered. This would require chemotherapy and/or radiotherapy prior to the transplant



Myeloma is normally treated with chemotherapy but a stem cell transplant 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. All diseases listed in the Haematology Appendix A, Classification are recordable for COSD



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 Leukaemia and Lymphoma 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

- 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