



National Cancer Institute

Hematopoietic and Lymphoid Neoplasm Project





Introduction to the WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues 4th edition



Hematopoietic and Lymphoid Lineages, Part II

Steven Peace, CTR

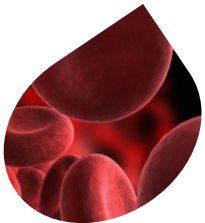
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September 2009



Objectives

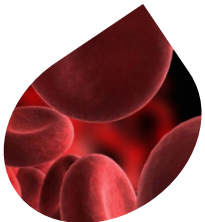
- Introduce Appendix B - 2010 Hematopoietic and Lymphoid Neoplasm Lineage Tables
- Understand when and how to use the 2010 Hematopoietic and Lymphoid Lineage Tables





2010 Hematopoietic and Lymphoid Tissue Lineage Tables

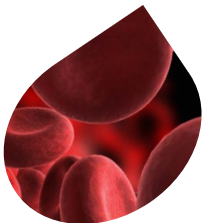
- Appendix B of the Manual
- When to Use Lineage Tables
- How to Determine Which Table to Use
 - *Only the WHO Preferred Terms are listed*
 - *Use Hematopoietic Database to identify synonyms that correspond to the WHO Preferred Term*
- How to Use the Individual Tables
- What is to be Included in Text Documentation





Appendix B – Histology Lineage

- 12 Tables – 6 myeloid/6 lymphoid
- WHO Classification Tables and 2010 Hematopoietic and Lymphoid Tissue Lineage Tables are IDENTICAL for malignant conditions
- WHO Tables include borderline conditions not of immediate interest as reportable diseases

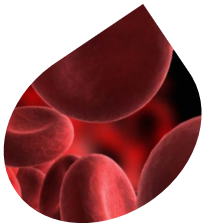


Appendix B – Histology Lineage

Appendix B WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues Histology Lineage

Note: Only the WHO Preferred Terms are listed.

Use the Hematopoietic Database to identify synonyms that correspond to the WHO Preferred Term.



Use of WHO Preferred Term

Appendix F Master Code Lists

Table F1a: WHO/ICD-O-3 Master List of Histology Codes – Alphabetical List

New 2010:	New histology terms and codes published in the <i>WHO Classification of Tumours of Hematopoietic and Lymphoid Tissues</i> , 4 th Edition. These codes are not in ICD-O-3 or the ICD-O-3 errata.
ICD-O-3 ONLY:	The histology term and code are published only in ICD-O-3
WHO and ICD-O-3:	The histology term is the WHO preferred term for that neoplasm; the WHO and ICD-O-3 preferred histology terms may not be the same, but the codes for this neoplasm are published in both WHO and in ICD-O-3.

Histologic Term	New 2010	ICD-O-3 Only	WHO and ICD-O-3
Acute basophilic leukemia			9870/3
Acute biphenotypic leukemia		9805/3	
Acute erythroid leukemia			9840/3
Acute megakaryoblastic leukemia			9910/3
Acute monoblastic and monocytic leukemia			9891/3
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13);RBM15-MKL1	9911/3		
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22), CBFβ/MYH11			9871/3
Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26.2);RPN1-EVI1	9869/3		
Acute myeloid leukemia with maturation			9874/3
Acute myeloid leukemia with minimal differentiation			9872/3
Acute myeloid leukemia with mutated CEBPA			9861/3
Acute myeloid leukemia with mutated NPM1			9861/3
Acute myeloid leukemia with myelodysplasia-related changes			9895/3
Acute myeloid leukemia with t(6;9)(p23;q34); DEK-NUP214	9865/3		

Use of WHO Preferred Term (2)

Table F1b: WHO/ICD-O-3 Master List of Histology Codes – Numerical List

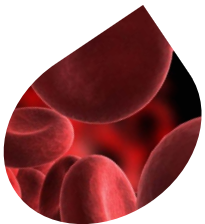
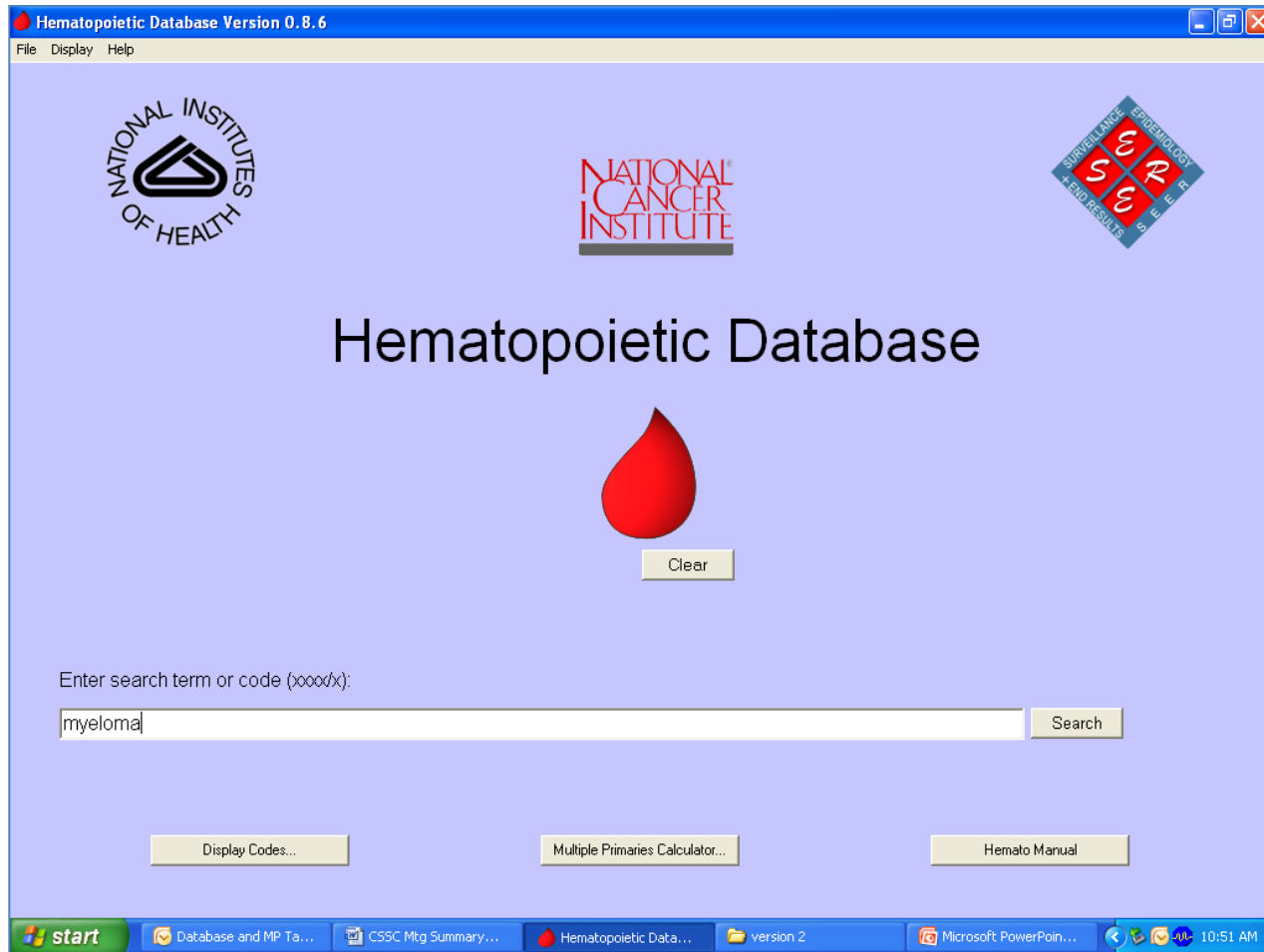
New 2010: New histology terms and codes published in the *WHO Classification of Tumours of Hematopoietic and Lymphoid Tissues*, 4th Edition. These codes are not in ICD-O-3 or the ICD-O-3 errata.

ICD-O-3 ONLY: The histology term and code are published only in ICD-O-3

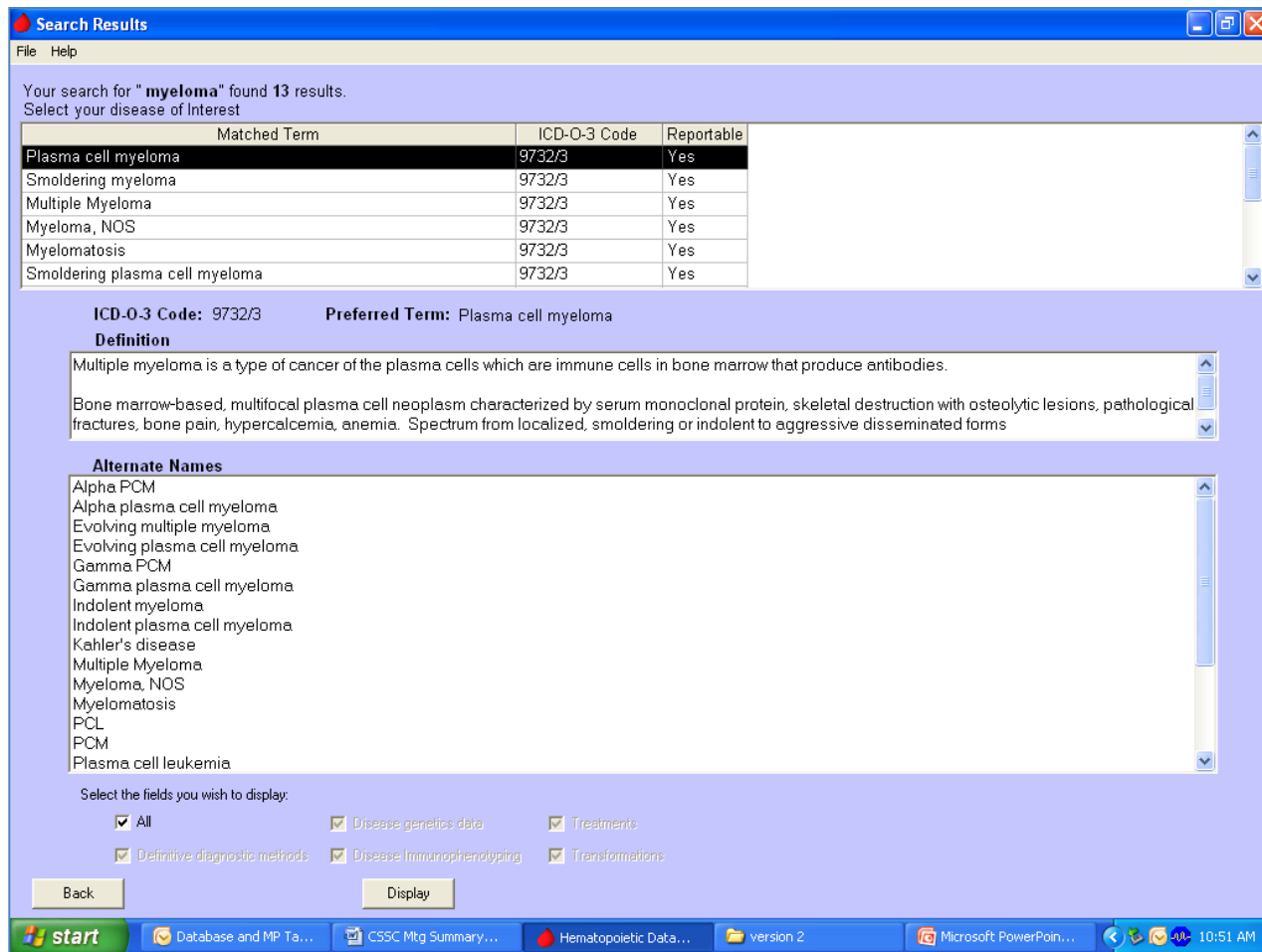
WHO and ICD-O-3: The histology term is the WHO preferred term for that neoplasm; the WHO and ICD-O-3 preferred histology terms may not be the same, but the codes for this neoplasm are published in both WHO and in ICD-O-3.

Histologic Term	New 2010	ICD-O-3 Only	WHO and ICD-O-3
Malignant lymphoma, NOS		9590/3	
Hairy cell leukemia-variant			9591/3
Splenic B-cell lymphoma/leukemia, unclassifiable			9591/3
Splenic diffuse red pulp small B-cell lymphoma			9591/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma			9596/3
Primary cutaneous follicle centre lymphoma	9597/3		
Classical Hodgkin lymphoma			9650/3
Lymphocyte-rich classical Hodgkin lymphoma			9651/3
Mixed cellularity classical Hodgkin lymphoma			9652/3
Lymphocyte-depleted classical Hodgkin lymphoma			9653/3
Hodgkin lymphoma, lymphocyte depletion, diffuse fibrosis		9654/3	
Hodgkin lymphoma, lymphocyte depletion, reticular		9655/3	
Nodular lymphocyte predominant Hodgkin lymphoma			9659/3
Hodgkin granuloma [obs]		9661/3	
Hodgkin sarcoma [obs]		9662/3	
Nodular sclerosis classical Hodgkin lymphoma			9663/3

Using Hematopoietic Database



WHO Preferred Term / Synonyms



Your search for "myeloma" found 13 results.
Select your disease of interest

Matched Term	ICD-O-3 Code	Reportable
Plasma cell myeloma	9732/3	Yes
Smoldering myeloma	9732/3	Yes
Multiple Myeloma	9732/3	Yes
Myeloma, NOS	9732/3	Yes
Myelomatosis	9732/3	Yes
Smoldering plasma cell myeloma	9732/3	Yes

ICD-O-3 Code: 9732/3 Preferred Term: Plasma cell myeloma

Definition

Multiple myeloma is a type of cancer of the plasma cells which are immune cells in bone marrow that produce antibodies.

Bone marrow-based, multifocal plasma cell neoplasm characterized by serum monoclonal protein, skeletal destruction with osteolytic lesions, pathological fractures, bone pain, hypercalcemia, anemia. Spectrum from localized, smoldering or indolent to aggressive disseminated forms

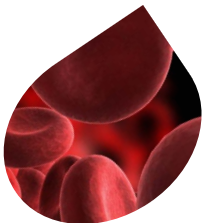
Alternate Names

- Alpha PCM
- Alpha plasma cell myeloma
- Evolving multiple myeloma
- Evolving plasma cell myeloma
- Gamma PCM
- Gamma plasma cell myeloma
- Indolent myeloma
- Indolent plasma cell myeloma
- Kähler's disease
- Multiple Myeloma
- Myeloma, NOS
- Myelomatosis
- PCL
- PCM
- Plasma cell leukemia

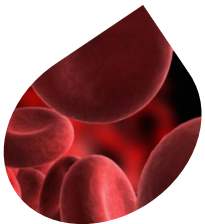
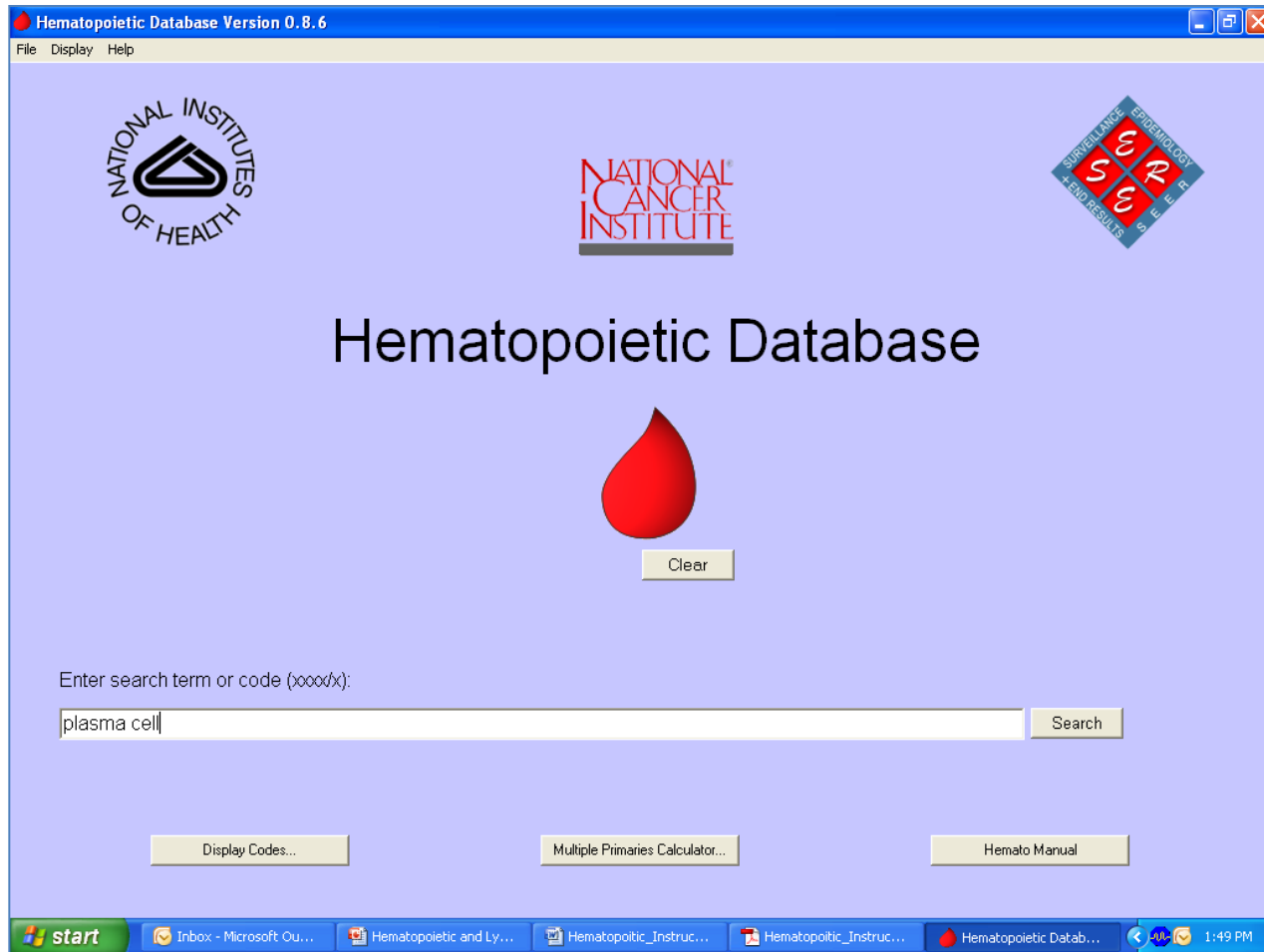
Select the fields you wish to display:

- All
- Disease genetics data
- Treatments
- Definitive diagnostic methods
- Disease Immunophenotyping
- Transformations

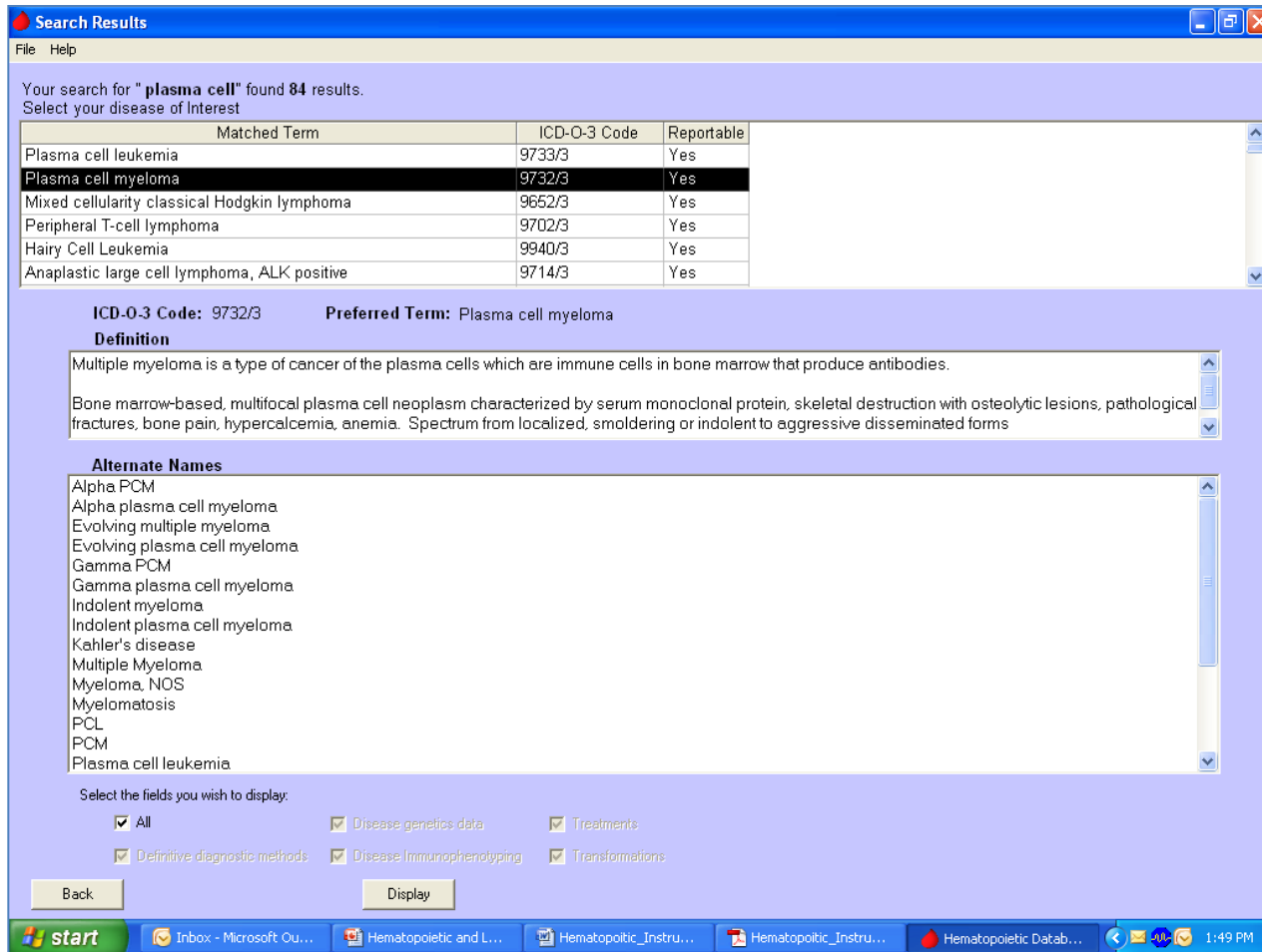
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Using Hematopoietic Database



WHO Preferred Term / Synonyms



Search Results

Your search for "plasma cell" found **84** results.
Select your disease of interest

Matched Term	ICD-O-3 Code	Reportable
Plasma cell leukemia	9733/3	Yes
Plasma cell myeloma	9732/3	Yes
Mixed cellularity classical Hodgkin lymphoma	9652/3	Yes
Peripheral T-cell lymphoma	9702/3	Yes
Hairy Cell Leukemia	9940/3	Yes
Anaplastic large cell lymphoma, ALK positive	9714/3	Yes

ICD-O-3 Code: 9732/3 **Preferred Term:** Plasma cell myeloma

Definition

Multiple myeloma is a type of cancer of the plasma cells which are immune cells in bone marrow that produce antibodies.

Bone marrow-based, multifocal plasma cell neoplasm characterized by serum monoclonal protein, skeletal destruction with osteolytic lesions, pathological fractures, bone pain, hypercalcemia, anemia. Spectrum from localized, smoldering or indolent to aggressive disseminated forms

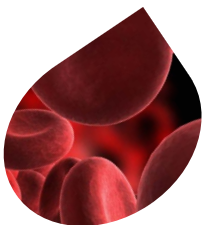
Alternate Names

- Alpha PCM
- Alpha plasma cell myeloma
- Evolving multiple myeloma
- Evolving plasma cell myeloma
- Gamma PCM
- Gamma plasma cell myeloma
- Indolent myeloma
- Indolent plasma cell myeloma
- Kähler's disease
- Multiple Myeloma
- Myeloma, NOS
- Myelomatosis
- PCL
- PCM
- Plasma cell leukemia

Select the fields you wish to display:

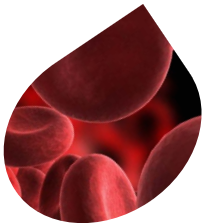
- All
- Disease genetics data
- Treatments
- Definitive diagnostic methods
- Disease Immunophenotyping
- Transformations

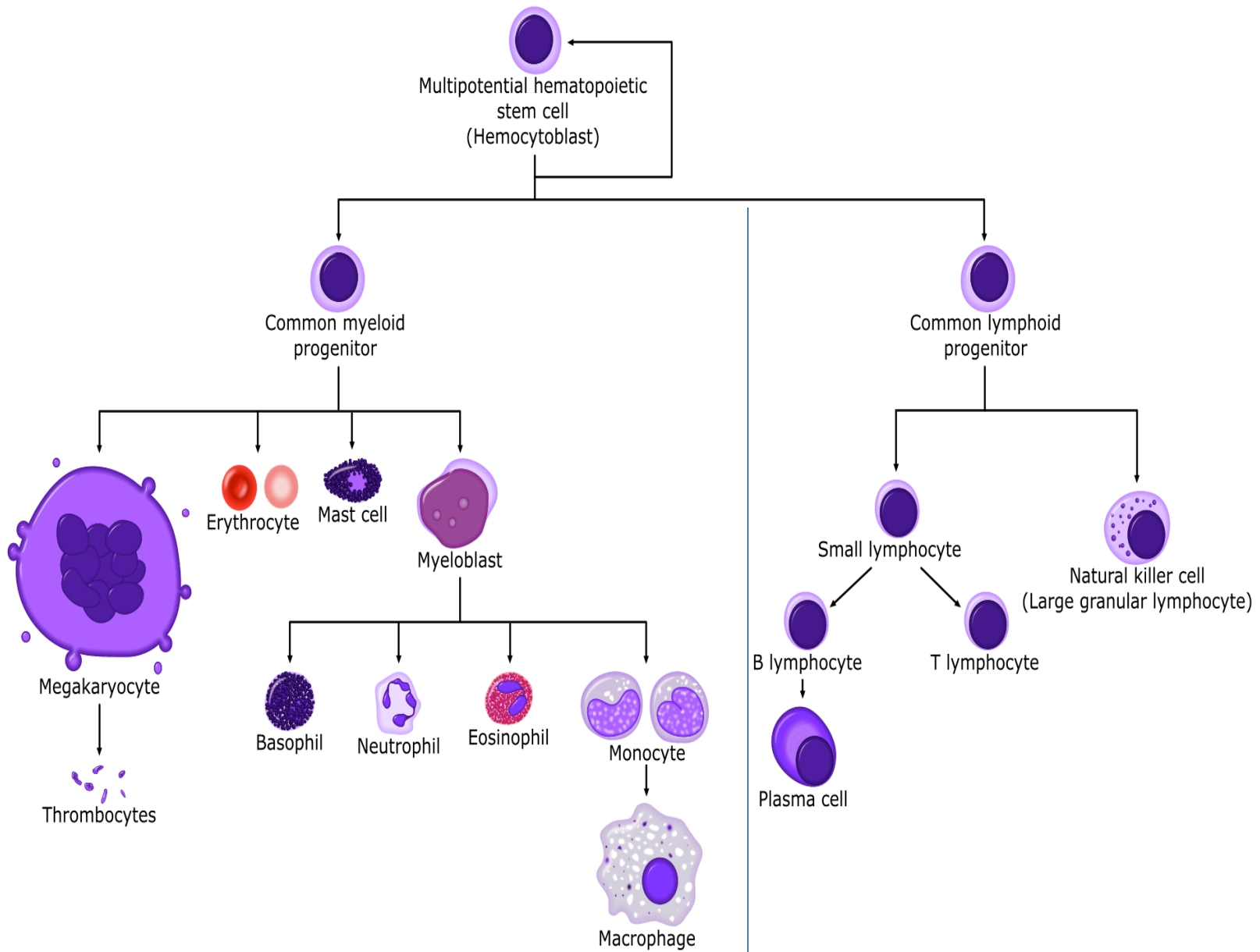
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Myeloid Line – 6 Tables

- Myeloproliferative Neoplasms
- Myeloid and Lymphoid Neoplasms with Eosinophilia and Abnormalities of PDGFRA, PDGFRB or FGFR1
- Myelodysplastic/Myeloproliferative Neoplasms
- Myelodysplastic Syndromes
- Acute Myeloid Leukemia and Related Precursor Neoplasms
- Acute Leukemias of Ambiguous Lineage



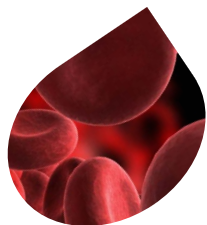


Hematopoietic stem cells give rise to two major progenitor cell lineages, myeloid and lymphoid progenitors
 Regenerative Medicine, 2006. <http://www.dentalarticles.com/images/hematopoiesis.png>

Myeloproliferative Neoplasms

Table B1: Myeloproliferative Neoplasms

WHO Preferred Term	ICD-O-3
Chronic eosinophilic leukemia, NOS	9964/3
Chronic myelogenous leukemia, BCR-ABL1 positive	9875/3
Chronic neutrophilic leukemia	9963/3
Cutaneous mastocytosis	9740/1
Essential thrombocythemia	9962/3
Mast cell leukemia	9742/3
Mast cell sarcoma	9740/3
Myeloproliferative neoplasm, unclassifiable	9975/3
Polycythemia vera	9950/3
Primary myelofibrosis	9961/3
Systemic mastocytosis	9741/3

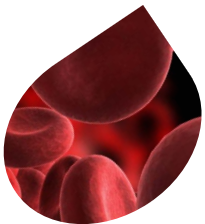




Myeloid and Lymphoid Neoplasms with Eosinophilia and Abnormalities of PDGFRA, PDGFRB or FGFR1

Table B2: Myeloid and Lymphoid Neoplasms with Eosinophilia and Abnormalities of PDGFRA, PDGFRB or FGFR1

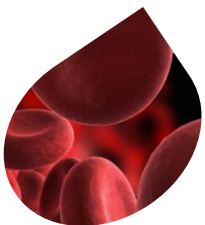
WHO Preferred Term	ICD-O-3
Myeloid and lymphoid neoplasm with FGFR1 abnormalities	9967/3
Myeloid and lymphoid neoplasm with PDGFRA rearrangement	9965/3
Myeloid neoplasm with PDGFRB rearrangement	9966/3



Myelodysplastic / Myeloproliferative Neoplasms

Table B3: Myelodysplastic / Myeloproliferative Neoplasms

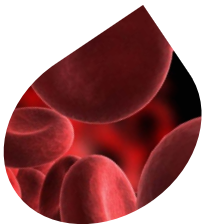
WHO Preferred Term	ICD-O-3
Atypical chronic myeloid leukemia, BCR-ABL1 negative	9876/3
Chronic myelomonocytic leukemia	9945/3
Juvenile myelomonocytic leukemia	9946/3
Myelodysplastic/myeloproliferative neoplasm, unclassifiable	9975/3
Refractory anemia with ring sideroblasts	9982/3



Myelodysplastic Syndromes

Table B4: Myelodysplastic Syndromes

WHO Preferred Term	ICD-O-3
Myelodysplastic syndrome associated with isolated del(5q)	9986/3
Myelodysplastic syndrome, unclassifiable	9989/3
Refractory anemia	9980/3
Refractory anemia with excess blasts	9983/3
Refractory anemia with ring sideroblasts	9982/3
Refractory cytopenia of childhood	9985/3
Refractory neutropenia	9991/3
Refractory thrombocytopenia	9992/3



Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms

Table B5: Acute Myeloid Leukemia (AML) and Related Precursor Neoplasms

WHO Preferred Term	ICD-O-3
Acute myeloid leukemias with recurrent genetic abnormalities	
Acute myeloid leukemia (megakaryoblastic) with t(1;22)(p13;q13); RBM15-MKL1	9911/3
Acute myeloid leukemia with inv(16)(p13.1q22) or t(16;16)(p13.1;q22); CBFβ-MYH11	9871/3
Acute myeloid leukemia with inv(3)(q21;q26.2) or t(3;3)(q21;q26.2); RPN1-EVI1	9869/3
Acute myeloid leukemia with t(8;21)t(q22;q22); RUNX1-RUNX1T1	9896/3
Acute myeloid leukemia with t(9;11)(p22;q23); MLLT3-MLL	9897/3
Acute promyelocytic leukemia (AML with t(15;17)(q22;q12), PML/RARA	9866/3
Acute myeloid leukemia with myelodysplasia-related changes	9895/3
Therapy-related myeloid neoplasm	9920/3

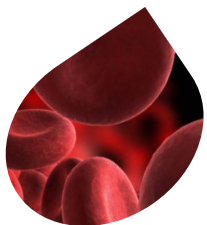
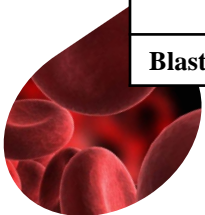


Table B5 continued

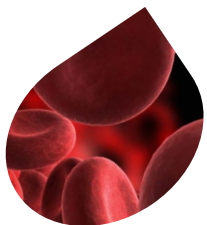
WHO Preferred Term	ICD-O-3
Acute myeloid leukemia, NOS	9861/3
Acute myeloid leukemia with minimal differentiation	9872/3
Acute myeloid leukemia without maturation	9873/3
Acute myeloid leukemia with maturation	9874/3
Acute myelomonocytic leukemia	9867/3
Acute erythroid leukemia	9840/3
Acute megakaryoblastic leukemia	9910/3
Acute basophilic leukemia	9870/3
Acute panmyelosis with myelofibrosis	9931/3
Myeloid sarcoma	9930/3
Myeloid proliferations related to Down syndrome	<i>No Code</i>
Transient abnormal myelopoiesis	9898/1
Myeloid leukemia associated with Down syndrome	9898/3
Blastic plasmacytoid dendritic cell neoplasm	9727/3



Acute Leukemias of Ambiguous Lineage

Table B6: Acute Leukemia of Ambiguous Lineage

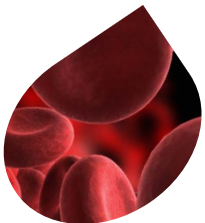
WHO Preferred Term	ICD-O-3
Acute undifferentiated leukemia	9801/3
Mixed phenotype acute leukemia with t(v;11q2); MLL rearranged	98073
Mixed phenotype acute leukemia, B/myeloid, NOS	9808/3
Mixed phenotype acute leukemia. T/myeloid, NOS	9809/3
Mixed phenotype acute leukemia with t(9;22)(q34;q11.2); BCR-ABL1	9806/3
Natural killer (NK) cell lymphoblastic leukemia/lymphoma	<i>No Code</i>

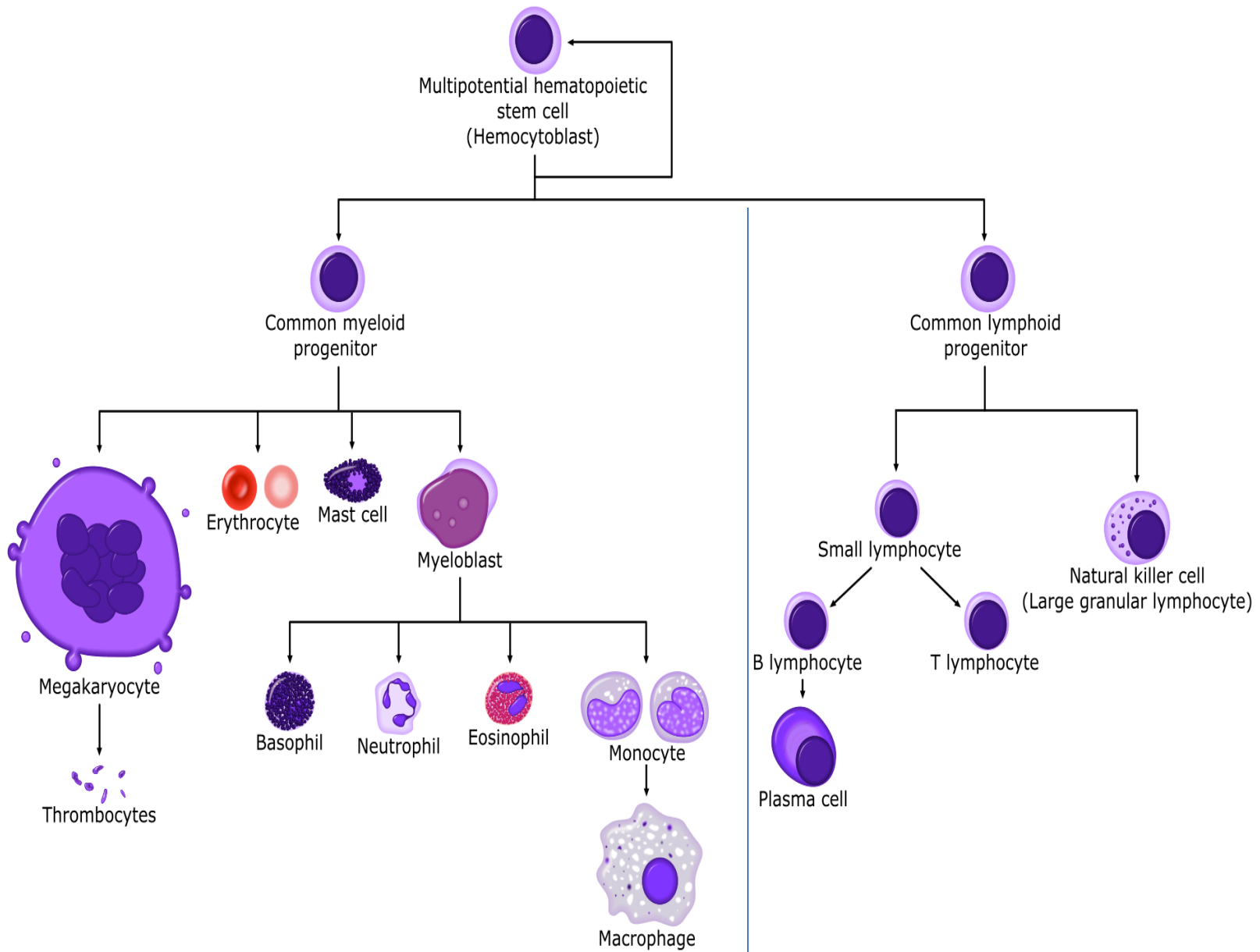




Lymphoid Line – 6 Tables

- Precursor Lymphoid Neoplasms
- Mature B-Cell Neoplasms
- Mature T-Cell and NK-Cell Neoplasms
- Hodgkin Lymphoma
- Histiocytic and Dendritic Cell Neoplasms
- Post-Transplant Lymphoproliferative Disorders



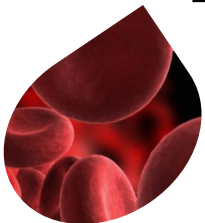


Hematopoietic stem cells give rise to two major progenitor cell lineages, myeloid and lymphoid progenitors
 Regenerative Medicine, 2006. <http://www.dentalarticles.com/images/hematopoiesis.png>

Precursor Lymphoid Neoplasms

Table B7: Precursor Lymphoid Neoplasms

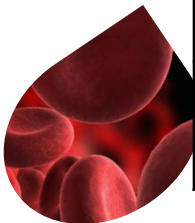
WHO Preferred Term	ICD-O-3
B lymphoblastic leukemia/lymphoma	<i>No Code</i>
B lymphoblastic leukemia/lymphoma with hyperdiploidy	9815/3
B lymphoblastic leukemia/lymphoma with hypodiploidy (hypodiploid ALL)	9816/3
B lymphoblastic leukemia/lymphoma with recurrent genetic abnormalities	<i>No Code</i>
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A-PBX1 (TCF3-PBX1)	9818/3
B lymphoblastic leukemia/lymphoma with t(12;21)(p13;q22); TEL-AML1 (ETV6-RUNX1)	9814/3
B lymphoblastic leukemia/lymphoma with t(5;14)(q31;q32); IL3-IGH	9817/3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812/3
B lymphoblastic leukemia/lymphoma with t(v;11q23); MLL rearranged	9813/3
B lymphoblastic leukemia/lymphoma, NOS	9811/3
T lymphoblastic leukemia/lymphoma	9837/3



Mature B-Cell Neoplasms – part I

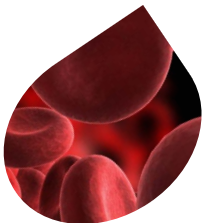
Table B8: Mature B-Cell Neoplasms

WHO Preferred Term	ICD-O-3
ALK positive large B-cell lymphoma	9737/3
B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and classical Hodgkin lymphoma	9596/3
B-cell prolymphocytic leukemia	9833/3
Burkitt lymphoma	9687/3
Chronic lymphocytic leukemia/small lymphocytic lymphoma	9823/3
Diffuse large B-cell lymphoma (DLBCL), NOS	9680/3
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)	9699/3
Extrasosseous plasmacytoma	9734/3
Follicular lymphoma	9690/3
Hairy cell leukemia	9940/3
Heavy chain disease	9762/3
Intravascular large B-cell lymphoma	9712/3



Mature B-Cell Neoplasms – part II

WHO Preferred Term	ICD-O-3
Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease	9738/3
Lymphomatoid granulomatosis	9766/1
Lymphoplasmacytic lymphoma	9671/3
Mantle cell lymphoma	9673/3
Plasma cell myeloma	9732/3
Plasmablastic lymphoma	9735/3
Primary cutaneous follicle centre lymphoma	9597/3
Primary effusion lymphoma	9678/3
Primary mediastinal (thymic) large B-cell lymphoma	9679/3
Solitary plasmacytoma of bone	9731/3
Splenic B-cell lymphoma/leukemia, unclassifiable	9591/3
Splenic marginal zone lymphoma	9689/3
T-cell/histiocyte rich large B-cell lymphoma	9688/3



Mature T-Cell and NK-Cell Neoplasms

Table B9: Mature T-Cell and NK-Cell Neoplasm

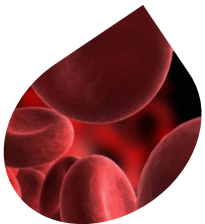
WHO Preferred Term	ICD-O-3
Adult T-cell leukemia/lymphoma	9827/3
Aggressive NK-cell leukemia	9948/3
Anaplastic large cell lymphoma, ALK positive	9714/3
Angioimmunoblastic T-cell lymphoma	9705/3
Chronic lymphoproliferative disorder of NK-cells	9831/3
Enteropathy-associated T-cell lymphoma	9717/3
Extranodal NK/T cell lymphoma, nasal type	9719/3
Hepatosplenic T-cell lymphoma	9716/3
Hydroa vacciniforme-like lymphoma	9725/3
Lymphomatoid papulosis	9718/3
Mycosis fungoides	9700/3
Peripheral T-cell lymphoma, NOS	9702/3
Primary cutaneous anaplastic large cell lymphoma	9718/3
Primary cutaneous CD30 positive T-cell lymphoproliferative disorders	No Code
Primary cutaneous CD4 positive small/medium cell T-cell lymphoma	9709/3
Primary cutaneous gamma-delta T-cell lymphoma	9726/3
Sezary syndrome	9701/3
Subcutaneous panniculitis-like T-cell lymphoma	9708/3
Systemic EBV positive T-cell lymphoproliferative disease of childhood	9724/3
T-cell prolymphocytic leukemia	9834/3



Hodgkin Lymphoma

Table B10: Hodgkin Lymphoma

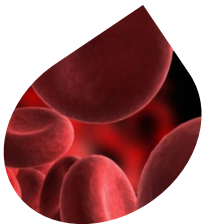
WHO Preferred Term	ICD-O-3
Classical Hodgkin lymphoma	9650/3
Lymphocyte-depleted classical Hodgkin lymphoma	9653/3
Lymphocyte-rich classical Hodgkin lymphoma	9651/3
Mixed cellularity classical Hodgkin lymphoma	9652/3
Nodular lymphocyte predominant Hodgkin lymphoma	9659/3
Nodular sclerosis classical Hodgkin lymphoma	9663/3



Histiocytic Cell and Dendritic Cell Neoplasms

Table B11: Histiocytic and Dendritic Cell Neoplasm

WHO Preferred Term	ICD-O-3
Disseminated juvenile xanthogranuloma	<i>No Code</i>
Fibroblastic reticular cell tumor	9759/3
Follicular dendritic cell sarcoma	9758/3
Histiocytic sarcoma	9755/3
Indeterminate dendritic cell tumor	9757/3
Langerhans cell histiocytosis	9751/3
Langerhans cell sarcoma	9756/3



Histiocytic Cell Neoplasms

Table 14.01 True histiocytic malignancy, a vanishing diagnosis.

Original diagnosis	Currently considered
Histiocytic lymphoma, nodular and diffuse	Diffuse large B-cell lymphoma Follicular lymphoma, grade 3 Peripheral T-cell lymphoma Histiocyte-rich variants of B-cell, T-cell, and Hodgkin lymphoma Anaplastic large cell lymphoma
Histiocytic medullary reticulosis	Haemophagocytic syndromes
Malignant histiocytosis	ALCL Haemophagocytic syndromes
Regressing atypical histiocytosis	Primary cutaneous CD30-positive T-cell lymphoma
Intestinal malignant histiocytosis	Enteropathy-type T-cell lymphoma
Histiocytic cytopathic panniculitis	Subcutaneous panniculitis-like T-cell lymphoma with Haemophagocytosis

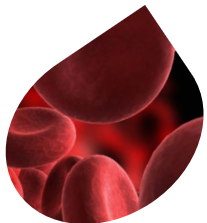


Post-Transplant Lymphoproliferative Disorders

Table B12: Post-Transplant Lymphoproliferative Disorders (PTLD)

WHO Preferred Term	ICD-O-3
Early lesions	<i>No Code</i>
Classical Hodgkin lymphoma type PTLD	*
Infectious mononucleosis-like PTLD	9971/1
Monomorphic PTLD (B- and T/NK-cell types)	*
Plasmacytic hyperplasia	9971/1
Polymorphic PTLD	9971/3

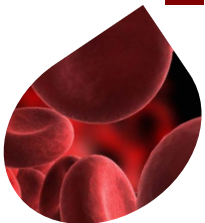
*These lesions are classified according to the leukemia or lymphoma to which they correspond, and are assigned the respective ICD-O code.





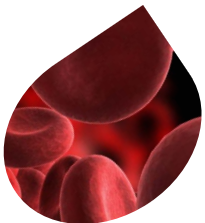
Module Review

- Hematopoiesis process
- Two primary hematopoietic cell lines
 - Myeloid
 - Lymphoid
- Lineages flow from the primary lines
- WHO Preferred Term and Synonyms
- Lineage Tables
 - 6 Myeloid cell line tables
 - 6 Lymphoid cell line tables
 - How and When to Use the Tables



References & More Information

- **WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues , 4th edition**, S. Swerdlow, E. Campo, N. Lee Harris, E. Jaffe, S. Pileri, H. Stein, J. Thiele, J. Vardiman, IAR C, Lyon, France, 2008
- **2010 Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual**, C. Hahn Johnson, M. Adamo, S. Peace, NCI SEER, 2009
- **Advances in Understanding and Management of Myeloproliferative Neoplasms**, Alessandro M. Vannucchi, Paola Guglielmelli and Ayalew Tefferi, *CA Cancer J Clin* 2009;59;171-191; Apr 15, 2009
- **Proposed Classification of Lymphoid Neoplasms for Epidemiologic Research from the International Lymphoma Epidemiology Consortium (Inter-Lymph)**, L. Morton, J. Turner, J. Cernan, *Blood*, DOI 10.1182/2006-11-0515672; Mar 27, 2007





Conclusion

- The new hematopoietic and lymphoid neoplasm rules go into effect for cases diagnosed **January 1, 2010, and after**
- Email address for questions askseerctr@imsweb.com

