

Abstracting and Coding Lymphoid Neoplasms

BACKGROUND MATERIAL
2014 HEMATOPOIETIC MANUAL AND DATABASE
CODING RULES AND INSTRUCTIONS
TREATMENT GUIDELINES
STAGING



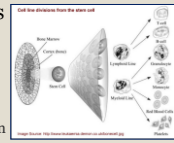
FCDS 2013-2014 Educational Webcast Series
Steven Peace, BS, CTR
February 20, 2014



Outline

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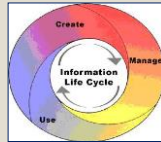
- Background and Characteristics
- Causes/Risk Factors/Signs/Symptoms
- Overview of the Immune System
- Hematopoiesis and Lymphoid Cell Line Derivation
- Anatomy of Two Circulatory Systems
- Complex Disease Processes
 - Confirming the Diagnosis
 - The Clinical Workup
 - Immunophenotype Studies
 - Identifying Disease Progression/Transformation



Outline

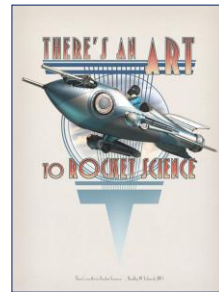
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- 2014 Updates to Tools & Rules
- Determining the Primary Site
- Determining the Histology
- Determining the Grade
- Staging Lymphoid Neoplasms
- Treatment for Lymphoid Neoplasms
- Text Documentation



Why Are These Cases So Challenging?

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Source: <http://shop.webomator.com/retropolis/prints/ArtToRocketScience.jpg>

Why Are These Cases So Challenging?

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- Not the same as when many of us started as registrars
- Terminology can be confusing and complicated
- Terms don't always match up with codes
- What is leukemia/lymphoma?
- Is multiple myeloma a type of leukemia?
- Are some lymphomas also leukemia and vice versa?
- Why are some lymphomas in lymph nodes but not all?



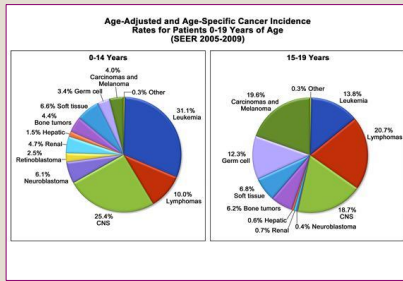
Inter-Lymph Classification Comparisons

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Real Classification	Proposed Classification	Working Formulation
B lymphoblastic	Preursor B lymphoblastic lymphoma/leukemia	Lymphoblastic
B lymphocytic, CLL	B cell chronic lymphocytic leukemia/leukemia	Small lymphocytic, consistent with CLL
B lymphocytic, postlymphocytic leukemia	postlymphocytic leukemia/leukemia	Small lymphocytic, plasmacytoid
Lymphoplasmacytoid immunocytoma	Lymphoplasmacytoid lymphoma	Small lymphocytic, plasmacytoid
Centrocytic	Mantle cell lymphoma	Diffuse, small cleaved cell
Centroblastic, centroblastic subtype		Follicular, small cleaved cell
		Diffuse, mixed small and large cell
		Diffuse, large cleaved cell
Centroblastic-centrocytic, follicular	Follicular center lymphoma, follicular	Follicular, predominantly small cleaved cell
	—Grade I	Follicular, mixed small and large cell
	—Grade II	Follicular, predominantly large cell
Centroblastic, follicular	Follicular center lymphoma, diffuse, small cell (provisional)	Diffuse, small cleaved cell
Centroblastic-centrocytic, diffuse		Diffuse, mixed small and large cell
		Diffuse, large cleaved cell
	Extranodal marginal zone B-cell lymphoma (non-nodal B-cell lymphoma of MALT type)	Small lymphocytic
	Nodal marginal zone B-cell lymphoma (provisional)	Diffuse, mixed small and large cell
		Diffuse, small cleaved cell
		Diffuse, mixed small and large cell
		Unclassifiable
	Splenic marginal zone B-cell lymphoma (provisional)	Small lymphocytic
		Diffuse, small cleaved cell
Hairy cell leukemia	Hairy cell leukemia	—
Plasmacytic	Plasmacytoma/plasmacytoma	Extranodular plasmacytoma
Centroblastic (monoclononic, polymorphic and multilobated subtypes)	Diffuse large B-cell lymphoma	Diffuse, large cell
		Large cell immunoblastic
Immunoblastic		Diffuse, mixed small and large cell
T-hive cell anaplastic, DLCL1		

Pediatric Neoplasms

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Source: NCI SEER Program

Adult Neoplasms

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Leading New Cancer Cases and Deaths - 2013 Estimates

Estimated New Cases*		Estimated Deaths	
Male	Female	Male	Female
Prostate	Breast	Lung & bronchus	Lung & bronchus
218,900 (28%)	232,860 (29%)	87,020 (28%)	72,220 (26%)
Lung & bronchus	Lung & bronchus	Prostate	Breast
116,000 (14%)	110,110 (14%)	29,220 (10%)	39,620 (14%)
Colon & rectum	Colon & rectum	Colon & rectum	Colon & rectum
73,680 (9%)	69,140 (9%)	26,300 (9%)	24,330 (9%)
Urinary bladder	Uterine corpus	Pancreas	Pancreas
54,650 (7%)	49,560 (6%)	18,980 (7%)	18,980 (7%)
Melanoma of the skin	Thyroid	Liver & intrahepatic bile duct	Chlary
45,000 (5%)	45,310 (6%)	14,500 (5%)	14,500 (5%)
Kidney & renal pelvis	Non-Hodgkin lymphoma	Leukemia	Leukemia
30,430 (4%)	30,430 (4%)	17,600 (6%)	10,600 (4%)
Non-Hodgkin lymphoma	Melanoma of the skin	Esophagus	Non-Hodgkin lymphoma
22,620 (3%)	22,620 (3%)	12,220 (4%)	8,420 (3%)
Oral cavity & pharynx	Kidney & renal pelvis	Uterine corpus	Uterine corpus
24,720 (3%)	24,720 (3%)	10,620 (4%)	8,190 (3%)
Leukemia	Pancreas	Non-Hodgkin lymphoma	Liver & intrahepatic bile duct
27,880 (3%)	22,480 (3%)	10,590 (4%)	6,780 (2%)
Pancreas	Chlary	Kidney & renal pelvis	Brain & other nervous system
22,740 (3%)	22,740 (3%)	8,780 (3%)	6,150 (2%)
All sites	All sites	All sites	All sites
854,290 (100%)	816,260 (100%)	316,000 (100%)	273,480 (100%)

*Excludes basal and squamous cell skin cancers and in situ carcinoma except urinary bladder.

©2013, American Cancer Society, Inc., Surveillance Research

Source: American Cancer Society

WHO Definition

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- “B cell and T/NK cell neoplasms are **clonal tumors of mature and immature B cells, T cells or natural killer (NK) cells** at various stages of differentiation.”
- Cells can be circulating lymphocytes such as lymphoid leukemia or cells in aggregate similar to a solid tumor but tumor made up of all the same type of cells (lymphoma).
- Features of clonality are most often used to identify and establish histologic type for most lymphoid neoplasm.

Lymphoid Neoplasm Characteristics

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- **2013 estimates in the United States**
 - 79,030 new lymphoma cases
 - ✦ 9,290 Hodgkin Lymphoma
 - ✦ 69,740 Non-Hodgkin Lymphoma
 - 20,200 lymphoma deaths
 - ✦ 1,180 Hodgkin Lymphoma Deaths
 - ✦ 19,020 Non-Hodgkin Lymphoma Deaths
- **2013 estimates in Florida**
 - 5,060 Non-Hodgkin Lymphoma Cases
 - 1,450 Non-Hodgkin Lymphoma Deaths



Source: American Cancer Society Cancer Facts and Figures 2013

Lymphoid Neoplasm Characteristics

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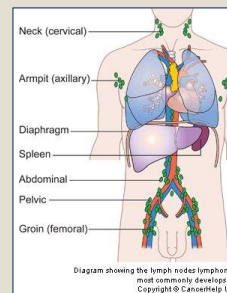
- **2013 estimates in the United States**
 - 15,680 Chronic Lymphocytic Leukemia
 - ✦ 4,580 CLL Deaths
 - 6,070 Acute Lymphocytic Leukemia
 - ✦ 1,430 ALL Deaths
- **2013 estimates in Florida**
 - 3,490 Leukemia Deaths
 - ✦ Lymphoid – CLL and ALL
 - ✦ Myeloid – CML and AML



Source: American Cancer Society Cancer Facts and Figures 2013

Common Lymph Node Chains for Lymphoma

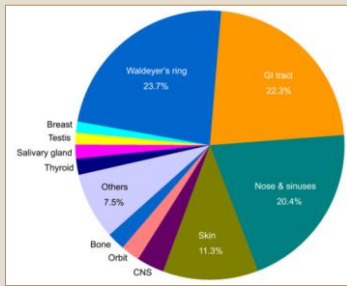
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Source: CancerHelpUK.org

Extra-Nodal Lymphoma

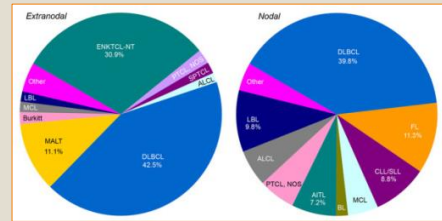
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Source: nlm.nih.gov

Common Types of Lymphoma

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Source: nlm.nih.gov

Causes and Risk Factors

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- Genetic Abnormalities (inherited/acquired)
- Conditions Causing Lowered Immunity
- Chemicals Causing Lowered Immunity
- History of Organ Transplant
- History of Viral or Bacterial Infection
 - HTLV1/HIV/EBV/HHV8/HepC/Helicobacter Pylori
- Auto Immune Condition
 - Rheumatoid Arthritis
 - Systemic Lupus Erythematosus
- Family History of Lymphoma

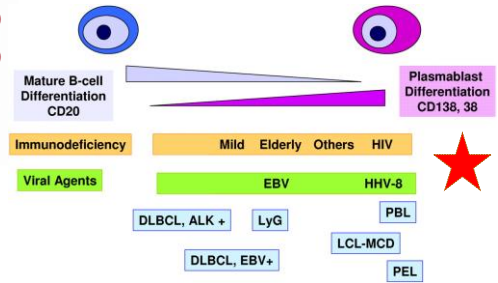


<http://cancer.gov/>

blood

JOURNAL OF THE AMERICAN SOCIETY OF HEMATOLOGY

Large B-cell lymphomas with a phenotype of terminal B-cell differentiation.



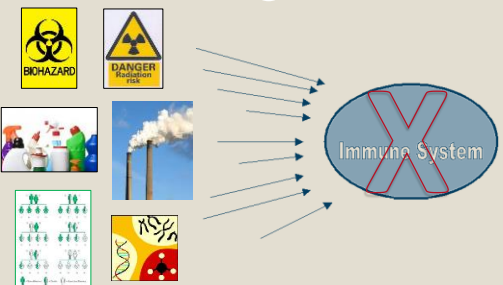
Campo E et al. Blood 2011;117:5019-5032

©2011 by American Society of Hematology

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Causes and Risk Factors

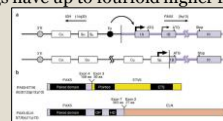
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Gene Mutation in Familial ALL

18

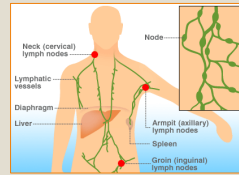
- Precursor B cell Acute Lymphoblastic Leukemia (pre-B ALL)
 - The most common malignancy in pediatrics
- PAX5 gene mutation or BSAP – inherited genetic mutation
- Mutated PAX5 present in 30% of pre-B ALL
- Genetic Alteration is 9p deletion with loss of heterozygosity (9p13)
- Identified as harbinger of germline mutation leading to pre-B ALL
- Affected siblings have up to fourfold higher risk for disease



Signs and Symptoms

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- Enlarged Lymph Node(s)
 - Neck
 - Armpit
 - Groin
- Swollen Abdomen
- Chest Pain/Pressure
- Shortness of Breath
- Fever
- Weight Loss
- Night Sweats
- Fatigue



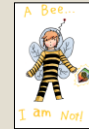
Source: b4tea.com

“B” Symptoms

20

- What is Significance of “B” Symptoms

- What are “B” Symptoms
 - Fevers
 - Night Sweats
 - Weight Loss > 10% of Body Weight
- Minor Symptoms
 - Malaise
 - Fatigue
 - Pruritis
 - Alcohol Intolerance
 - Frequent Infections



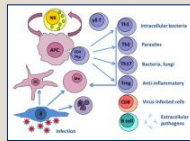
Not a “B”

- **Do Not Code Minor Symptoms as “B” Symptoms**

Immune System

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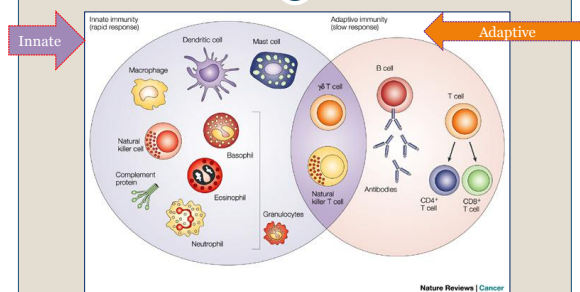
- Primary Function Lymphatic System – Fluid Retrieval
- Primary Function Immune System – Protect from infection
 - Bacteria
 - Viruses
 - Fungi
 - Injury
 - Parasites
- Interacts with Nervous System
- Protects via immune response from:
 - Innate Immunity
 - Adaptive Immunity



Source: <http://static.abdsrotec.com/2013images/figure1.jpg>

Immune System

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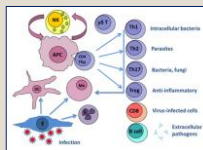


Source: Nature Reviews/Cancer

Immune System

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- Causes of Lymph Node Enlargement:
 - Non-specific reactive hyperplasia
 - Inflammatory Reaction
 - Foreign Body
 - Tuberculosis
 - Infection
 - Injury
 - Neoplasm
 - Primary – Lymphoma (Hodgkin or Non-Hodgkin)
 - Secondary – Metastatic Ds. via Lymph Node Drainage

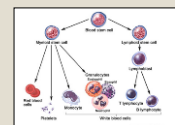


Source: <http://static.abdsrotec.com/2013images/figure1.jpg>

Hematopoiesis

24

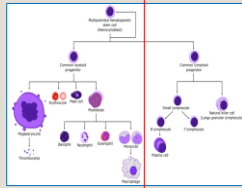
- What is a hematopoietic stem cell?
- Where are hematopoietic stem cells found?
- Hematopoietic stem cells give rise to ALL blood cells in a process called Cell Line Differentiation
 - Lymphoid cell line (lineage)
 - Myeloid cell line (lineage)
- Cell Line Differentiation
- Cell Line Proliferation
- Regulating Proliferation and Differentiation



Regulatory Function of Cells

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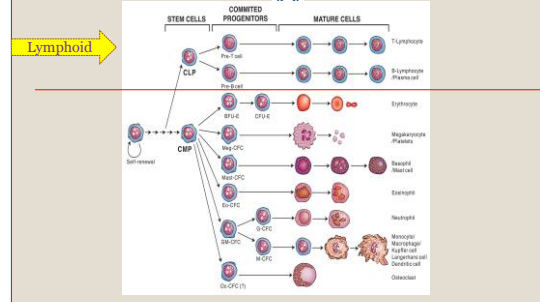
- Regulation of proliferation
- Regulation of differentiation
- Turn on/Turn off
 - Growth factors
 - Genes (including mutations)
 - Proteins
- Disregulation disrupts normal development of cell line
- Oncogenesis – becoming malignant



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

Lymphoid Cell Line Differentiation

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Blood Lines – Donald Metzger, AlphaMD Press, 2005
Figure 3.2 The eight major hematopoietic lineages generated by self-renewing multipotential stem cells
Copyright © 2008 by AlphaMed Press

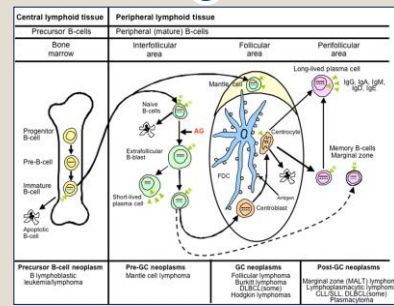
Cell Line Differentiation

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- Cellular differentiation is the process by which an immature cell becomes a more mature cell
- Differentiation changes a cell's size, shape, membrane potential, metabolic activity, and responsiveness to signals or signal pathways
- Regulatory function of cells (regulates cell line proliferation and cell line differentiation) so you have right mix of different types of hematopoietic cells being produced by the bone marrow...and circulating in the blood and/or lymph.
- Over/Under Production by bone marrow of one cell line (clonal)
- Too many or too few cells may lead to chronic/acute condition

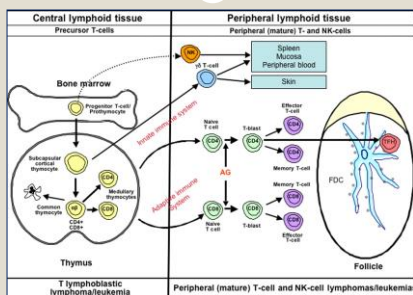
B-cell Differentiation

28



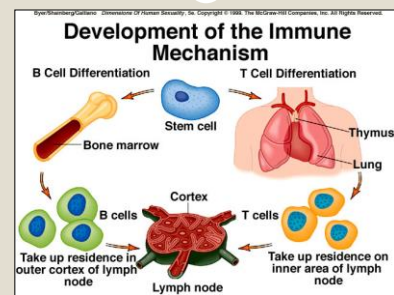
T-cell Differentiation

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Differentiation and Immune Function

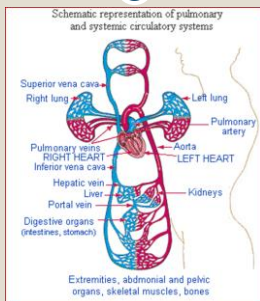
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Source: www.mhhe.com/science

Blood Circulatory System

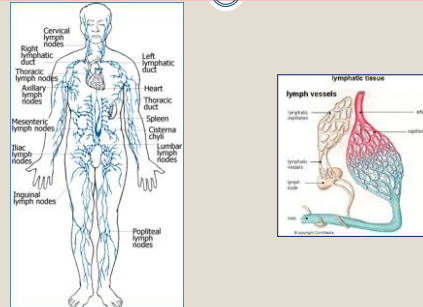
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Source: <http://webschoolsolutions.com/patts/systems/heart.htm>

Lymphatic Circulatory System

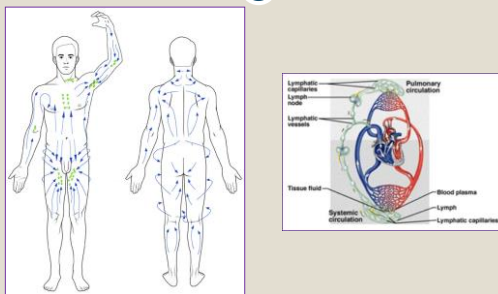
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Source: http://www.gorhams.dk/html/the_lymphatic_system.htm

Lymphatic Circulatory System

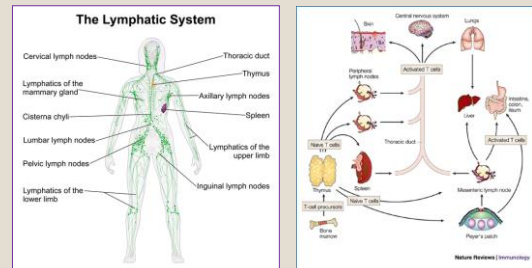
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Source: Nature Reviews Immunology <http://www.nature.com/nri/journal/v4/n5>

The Lymphatic System

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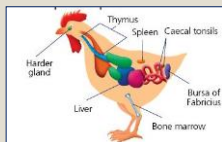


Lymphatic Organs

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Primary Organs

- Bone Marrow
- Thymus



Secondary Organs

- Spleen – process blood
 - Red Pulp
 - White Pulp
- Tonsils (Waldeyer's Ring)
- Lymph Nodes – process extracellular fluids
- MALT (mucosa-associated lymphoid tissue) – process mucosa
 - GALT (gut-associated lymphoid tissue)
 - Peyer's Patches
- Skin

Lymphatic Organs

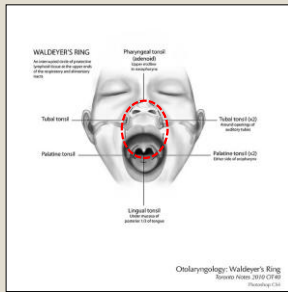
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<http://commonsensehealth.com>

Lymphatic Organs

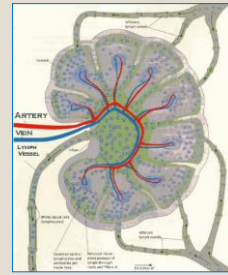
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http://www.flickr.com/photos

Lymph Node

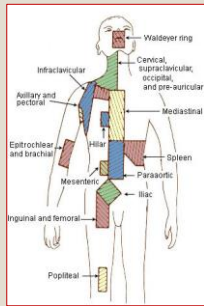
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Source: http://www.bcb.uwc.ac.za/SC_ED/grade10/manphys/plan.htm

Lymph Node Chains

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Lymph nodes above the diaphragm

1. Waldeyer's ring
2. Cervical, supraclavicular, occipital, and pre-auricular
3. Infraclavicular
4. Axillary and pectoral
5. Mediastinal
6. Hilar
7. Epitrochlear and brachial

Lymph nodes below the diaphragm

8. Spleen
9. Mesenteric
10. Para-aortic
11. Iliac
12. Inguinal and femoral
13. Popliteal

Source: AJCC Cancer Staging Form, 7th edition

2012 Hematopoietic and Lymphoid Neoplasm Core Reproductibility and Coding Manual

Appendix C Lymph Node/Lymph Node Chain Reference Table

Use this table with the Primary Site and Histology Rules to determine whether involved lymph nodes are in a single ICD-O-3 lymph node region or in multiple ICD-O-3 lymph node regions.
This table contains the names of lymph nodes that have the capsule and sinus structure of true lymph nodes. Lymphoid tissue such as that in the GI tract, tonsils, etc. is not represented in this table.
Note: Pathologic reports may identify lymph nodes within most organs; the most common being breast, parotid gland, lung, and prostate. The lymph nodes in these organs are called extra-organ nodes; lymph nodes such as metastatic lymph nodes. We have included the most common extra-organ lymph nodes in this table. For an intra-organ lymph node not listed on the table, code to the ICD-O-3 topography code for that organ's regional lymph node (if any).

Table C1: Lymph Node/Lymph Node Chain Reference Table

Lymph Node/Lymph Node Chain	Use for MPII	ICD-O-3 Lymph Node Region(s)	AJCCCS Staging
Abdominal	C72	Intra-abdominal	Mesenteric
Axillary	C42	Pitc	Pitc; right and left*
Anterior axillary	C42	Axilla or arm	Axillary; right and left*
Anterior axil	C42	Intra-abdominal	Mesenteric
Anterior deep cervical	C70	Head, face and neck	Cervical; right and left*
Anterior jugular	C70	Head, face and neck	Cervical; right and left*
Aortic NOS, ascending aortic; lateral aortic; lumbar aortic; para-aortic; post-aortic	C72	Intra-abdominal	Para-aortic
Aortic NOS, ascending aortic window (subcostal)	C72	Intra-abdominal	Para-aortic
Appendicular	C72	Intra-abdominal	Mesenteric
Ascending aortic	C72	Intra-abdominal	Para-aortic
Axilla, 1/axilla (nodes near axilla)	C42	Intra-abdominal	Para-aortic
Anterior NOS, infra-axillary; pre-axillary; post-axillary; retro-axillary	C70	Head, face and neck	Cervical; right and left*
Axillary, lateral	C42	Axilla or arm	Axillary; right and left*
Axillary, anterior	C42	Axilla or arm	Axillary; right and left*
Azygos (lower paratracheal)	C72	Intra-abdominal	Mediastinal
Brachial	C72	Head or arm	Cervical; right and left*
Brachiocephalic; brachiocephalic; hilar; preaxillary; pulmonary root	C72	Intra-abdominal	Hilar
Brachiocephalic; bronchopulmonary; bronchial hilar; preaxillary; pulmonary root	C72	Intra-abdominal	Hilar
Brachiocephalic; bronchopulmonary; bronchial hilar; preaxillary; pulmonary root	C72	Intra-abdominal	Hilar
Brachial	C70	Head, face and neck	Cervical; right and left*
Brachiocephalic (axillary)	C42	Head, face and neck	Cervical; right and left*
Clavicular node (scroto-lymphic triangle or lympho-hilary triangle)	C72	Intra-abdominal	Para-aortic
Clavicular	C72	Intra-abdominal	Mediastinal

Version 2.2 (February 2012)

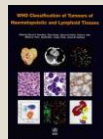
Effective with Code Disposition 1.1.2012 and after

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Classification of Lymphoid Neoplasms

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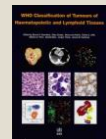
- Development of a World Standard
 - 1951 – Dameshek – clinical phenotype
 - 1960 – Philadelphia (Ph1) chromosome
 - 1966 – Rappaport Classification
 - 1974 – Kiel Classification System
 - 1974 – Lukes and Collins System
 - 1976 – Revised Rappaport Classification
 - 1976 – French – American – British Classification



Classification of Lymphoid Neoplasms

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- Development of a World Standard
 - 1982 – Working Formulation
 - 1994 – Revised European-American Classification of Lymphoid Neoplasms
 - 2001 – WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues, 3rd edition, 2001
 - 2008 – WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues, 4th edition, October 2008

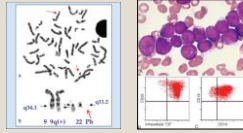
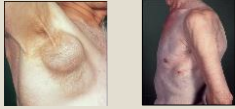
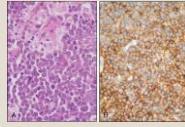


2008 WHO Classification of Lymphoid Neoplasms

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Incorporates:

- ✓ Histology/Morphology
- ✓ Stage of Differentiation
- ✓ Immunophenotype
- ✓ Genotypic features
- ✓ Clinical features



2008 WHO Classification - Lymphoid

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Table B7: Precursor Lymphoid Neoplasms

Precursor Lymphoid Neoplasm

WHO Preferred Term	Code
Adult T-cell leukemia/lymphoma	9817.3
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	No Code
B lymphoblastic leukemia/lymphoma with t(1;19)(q23;p13.3); E2A-PBX1 (TCF7-PBX1)	9818.3
B lymphoblastic leukemia/lymphoma with t(12;21)(q24;q22); TEL-AML1 (ETV6-RUNX1)	9814.3
B lymphoblastic leukemia/lymphoma with t(3;14)(q21;q21); ILL3-JGH	9817.3
B lymphoblastic leukemia/lymphoma with t(9;22)(q34;q11.2); BCR-ABL1	9812.3
B lymphoblastic leukemia/lymphoma with t(11;14)(q23); MLL rearranged	9813.3
B lymphoblastic leukemia/lymphoma, NOS	9811.3

Table B8: Mature B-Cell Neoplasms

Mature B-Cell Neoplasm

WHO Preferred Term	Code
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)	9680.3
Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)	9699.3
Extranodal plasmacytoma	9734.3
Follicular lymphoma	9690.3
Hairy cell leukemia	9940.3

2008 WHO Classification - Lymphoid

45

Mature B-Cell Neoplasm (con't)

WHO Preferred Term	Code
Heavy chain disease	9728.3
Intravascular large B-cell lymphoma	9729.3
Large B-cell lymphoma arising in HIV-associated multicentric Castlemans disease	9730.3
Lymphomatous meningitis	9661.3
Lymphoplasmacytic lymphoma	9671.3
Mantle cell lymphoma	9675.3
Nim-Hodgkin lymphoma, NOS, variant: B-cell lymphoma/leukemia, unclassifiable	9593.3
Plasma cell dyscrasia	9751.3
Plasmablastic lymphoma	9752.3
Primary cutaneous follicle center lymphoma	9753.3
Primary cutaneous marginal zone lymphoma	9679.3
Primary diffuse large B-cell lymphoma	9678.3
Primary nodular lymphoma, large B-cell lymphoma	9679.3
Splenic plasmacytoma of bone	9754.3
Splenic marginal zone lymphoma	9689.3
T-cell/histiocytic-rich large B-cell lymphoma	9683.3
Waldenström Macroglobulinemia	9761.3

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

Mature T-Cell and NK-Cell Neoplasm

WHO Preferred Term	Code
Adult T-cell leukemia/lymphoma (HTLV-1 positive)	9827.3
Anaplastic NK cell leukemia	9648.3
Anaplastic large cell lymphoma, ALK positive	9744.3
Anaplastic large cell lymphoma, ALK negative	9745.3
Anaplastic T-cell lymphoma	9746.3
Anaplastic T-cell lymphoma, variant type	9747.3
Hepatoerythrodermic T-cell lymphoma	9748.3
Histiocytic lymphoma-like lymphoma	9749.3
Lymphomatous panniculitis	9750.3
Subcutaneous panniculitis-like T-cell lymphoma	9755.3
Systemic T-cell lymphoma, NOS	9756.3
Systemic T-cell lymphoma, NOS	9757.3
Systemic T-cell lymphoma, NOS	9758.3
Systemic T-cell lymphoma, NOS	9759.3
Systemic T-cell lymphoma, NOS	9760.3
Systemic T-cell lymphoma, NOS	9761.3
Systemic T-cell lymphoma, NOS	9762.3
Systemic T-cell lymphoma, NOS	9763.3
Systemic T-cell lymphoma, NOS	9764.3
Systemic T-cell lymphoma, NOS	9765.3
Systemic T-cell lymphoma, NOS	9766.3
Systemic T-cell lymphoma, NOS	9767.3
Systemic T-cell lymphoma, NOS	9768.3
Systemic T-cell lymphoma, NOS	9769.3
Systemic T-cell lymphoma, NOS	9770.3
Systemic T-cell lymphoma, NOS	9771.3
Systemic T-cell lymphoma, NOS	9772.3
Systemic T-cell lymphoma, NOS	9773.3
Systemic T-cell lymphoma, NOS	9774.3
Systemic T-cell lymphoma, NOS	9775.3
Systemic T-cell lymphoma, NOS	9776.3
Systemic T-cell lymphoma, NOS	9777.3
Systemic T-cell lymphoma, NOS	9778.3
Systemic T-cell lymphoma, NOS	9779.3
Systemic T-cell lymphoma, NOS	9780.3
Systemic T-cell lymphoma, NOS	9781.3
Systemic T-cell lymphoma, NOS	9782.3
Systemic T-cell lymphoma, NOS	9783.3
Systemic T-cell lymphoma, NOS	9784.3
Systemic T-cell lymphoma, NOS	9785.3
Systemic T-cell lymphoma, NOS	9786.3
Systemic T-cell lymphoma, NOS	9787.3
Systemic T-cell lymphoma, NOS	9788.3
Systemic T-cell lymphoma, NOS	9789.3
Systemic T-cell lymphoma, NOS	9790.3
Systemic T-cell lymphoma, NOS	9791.3
Systemic T-cell lymphoma, NOS	9792.3
Systemic T-cell lymphoma, NOS	9793.3
Systemic T-cell lymphoma, NOS	9794.3
Systemic T-cell lymphoma, NOS	9795.3
Systemic T-cell lymphoma, NOS	9796.3
Systemic T-cell lymphoma, NOS	9797.3
Systemic T-cell lymphoma, NOS	9798.3
Systemic T-cell lymphoma, NOS	9799.3
Systemic T-cell lymphoma, NOS	9800.3

2008 WHO Classification - Lymphoid

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Hodgkin Lymphoma

WHO Preferred Term	Code
Classical Hodgkin lymphoma	9650.3
Lymphocyte-depleted classical Hodgkin lymphoma	9651.3
Lymphocyte-rich classical Hodgkin lymphoma	9652.3
Mixed cellularity classical Hodgkin lymphoma	9653.3
Nodular sclerosing classical Hodgkin lymphoma	9654.3

Table B11: Histiocytic and Dendritic Cell Neoplasms

Histiocytic/Dendritic Cell Neoplasm

WHO Preferred Term	Code
Disseminated gastric xanthogranuloma	No Code
Follicular dendritic cell tumor	9750.3
Follicular dendritic cell sarcoma	9751.3
Histiocytic sarcoma	9752.3
Immunoproliferative cell tumor	9757.3
Langerhans cell histiocytosis	9758.3
Langerhans cell sarcoma	9759.3

Table B12: Post-Transplant Lymphoproliferative Disorders (PTLD)

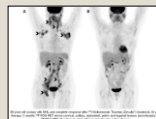
PTLD (Post-Transplant)

WHO Preferred Term	Code
EBV positive	9694.3
EBV negative	9695.3
Classical Hodgkin lymphoma type PTLD	-
Lymphomatous PTLD (B- and T/NK-cell types)	-
Plasmacytic hyperplasia	8091.3
Post-transplant lymphoproliferative disorder	9693.3

Understanding Complex Disease Processes

47

- Lymph Node Biopsy
- Extranodal Site Biopsy
- Diagnostic Imaging (CT/PET/MRI)
- Bone Marrow Aspirate
- Bone Marrow Biopsy

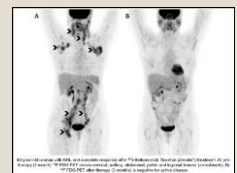


- Histology/Morphology
- Immunohistochemistry
- Flow Cytometry (Immunophenotype)
- Cytogenetics
- Molecular Genetic Studies
 - FISH
 - PCR

The Clinical Workup

48

- Disease Definition
- Risk Factors
- Signs and Symptoms
- Diagnostic Work Up
 - Clinical Evaluation
 - History and Physical
 - CBC – What is Normal
 - Immunophenotype
 - Imaging Studies (CT/PET/MRI and PET/CT)
 - Tissue Biopsy – Histologic Type and Staining
 - Bone Marrow Biopsy – Histologic Type and Staining
 - Molecular Cytogenetics – Genetic Testing



Disease Definition

49

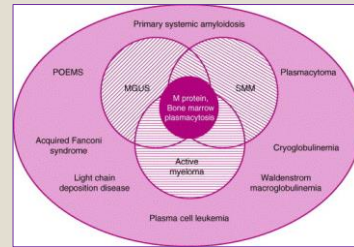
Table 2
Diagnostic criteria for plasma cell myeloma

Symptomatic plasma cell myeloma
M-protein in serum or urine ¹
BM clonal plasma cells or plasmacytoma ²
Related organ or tissue impairment heavy chain disease ³ (CRAB)
Asymptomatic (smoldering) myeloma
M-protein in serum at myeloma levels (> 30 g/L) and/or ≥ 10% clonal plasma cells in BM
No related organ or tissue impairment end-organ damage or bone lesions [CRAB] or myeloma-related symptoms

Source: BLOOD, 12 MAY 2011 VOLUME 117, NUMBER

Plasma Cell Neoplasms

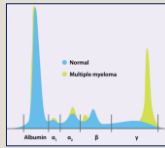
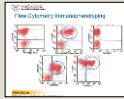
50



Immunophenotype

51

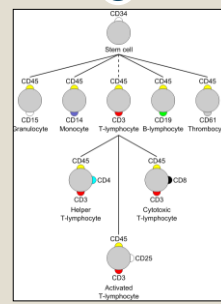
- Study of proteins expressed by cells
- Evaluates or Designates
 - Proliferation (myeloid or lymphoid)
 - Differentiation (category of malignancy)
- Antibodies "cluster of differentiation" or "CD"
- Immunophenotyping methods
 - Immunohistochemistry
 - Immunofluorescence
 - Flow cytometry
 - Electrophoresis



Source: <http://www.mayomedicallaboratories.com/articles/>

Cluster of Differentiation

52



Source: Schorschski @ de.wikipedia

Cluster of Differentiation Markers – B Cell

53

B-cell CD markers										
Marker Status	CD5	CD10	CD19	CD20	CD21	CD22	CD23	CD43	CD79a	slg
Follicular	1	3	4	4	4	2	1	4	4	-
Nodal marginal zone	1	1	4	4	4	1	2	4	M4, D1	-
MAIT	1	1	4	4	4	1	2	4	M4	-
Splenic Marginal zone	1	1	4	4	4	1	0	4	M4	-
CLL/SLL	4	0	4	4	4	4	4	4	D3	-
Lymphoplasmacytic Waldenström	1	1	4	4	4	0	3	4	M4, D2	-
Mantle Cell	4	1	4	4	4	1	4	4	M4, D4	-
Precursor B-cell (lymphoblastic)	4	3	4	4	4	0	0	4	0	-
Diffuse large B-cell	2	2	4	4	4	0	1	4	-	-
Mediastinal large cell	2	-	-	-	-	-	-	-	-	-
Burkitt's	1	4	-	-	-	-	-	-	-	-
Intravascular B-cell	-	-	-	-	-	-	-	-	-	-

Footnote: 0 = negative, 1 = <10% positive, 2 = 10-50% positivity, 3 = 50-90% positivity and 4 = >90% positive

Source: <http://www.nhlcberfamily.org/tests/cdmarkers.htm>

Cluster of Differentiation Markers – T Cell

54

T-cell CD markers									
Marker Status	CD3	CD5	CD7	CD4	CD8	CD30	NK16/56		
Type									
T-prolymphocytic leukaemia	+	+	+	+	(-)	-	-		
T-large granular lymphoproliferative	+	+	+	-	+	-	-		
Mycoid: Fungoides	+	+	+	+	(-)	(-)	-		
Cutaneous ALCL	+	(-)	(-)	(-)	+	++	(-)/(-)		
Primary systemic ALCL	(-)	(-)	(-)	(-)	(-)	++	-		
Peripheral T-cell lymphoma, unspecified	(-)	(-)	(-)	(-)	(-)	(-)	(-)/(-)		
Subcutaneous panniculitis-like T-cell	+	+	+	(-)	(-)	(-)	(-)		
Hepatosplenic T-cell lymphoma	+	+	+	-	-	-	+/(-)		
Angioimmunoblastic T-cell lymphoma	+	+	+	(-)	(-)	-	-		
Extranodal NK/Tcell lymphoma	S, C, +	-	(-)	(-)	-	-	-		
Eatropathy-associated T-cell lymphoma	+	+	+	(-)	(-)	(-)	-		
Adult T-cell leukaemia/lymphoma	+	+	+	(-)	(-)	(-)	-		

Footnote: + = >90% positive; (-) = >50% positive; () = <50% positive; - = <10% positive. ALCL-Anaplastic large cell lymphoma; C=Cytoplasmic; S-Surface.

Source: <http://www.nhlcberfamily.org/tests/cdmarkers.htm>

Dx Confirmation - Codes

55

Codes: Hematopoietic or Lymphoid Tumors (9590-9992)

Code	Description	Definition
1	Positive histology	Histologic confirmation (no tissue macroscopically examined).
2	Positive cytology	Cytologic confirmation (no tissue macroscopically examined, fluid cells macroscopically examined).
3	Positive histology PLUS • Positive immunophenotyping AND/OR • Positive genetic studies	Histology is positive for cancer, and there are also positive immunophenotyping and/or genetic test results. For example, bone marrow examination is positive for acute myeloid leukemia (M4) (J) (Genetic testing shows AML with inv(16)(p13;q22) (1971.3)).
4	Positive microscopic confirmation, method not specified	Microscopic confirmation is all that is known. It is unknown if the cells were from histology or cytology.
5	Positive laboratory test/markers study	A clinical diagnosis of cancer is based on laboratory test/markers studies which are clinically diagnostic for cancer.
6	Direct visualization without microscopic confirmation	The tumor was visualized during a surgical or endoscopic procedure only with no tissue resected for microscopic examination.
7	Radiography and other imaging techniques without microscopic confirmation	The malignancy was reported by the physician from an imaging technique report only.
8	Clinical diagnosis only, other than 5, 6 or 7	The malignancy was reported by the physician in the medical record.
9	Unknown whether or not macroscopically confirmed	A statement of malignancy was reported in the medical record, but there is no statement of how the cancer was diagnosed (usually accuracy).

Dx Confirmation - Instructions

56

Coding Instructions for Hematopoietic or Lymphoid Tumors (9590-9992)

- There is no priority hierarchy for coding *Diagnostic Confirmation* for hematopoietic and lymphoid tumors. Most commonly, the specific histologic type is diagnosed by immunophenotyping or genetic testing. See the *Hematopoietic Database (DB)* for information on the definitive diagnostic confirmation for specific types of tumors.
- Code 1 when the microscopic diagnosis is based on tissue specimens from biopsy, frozen section, surgery, or autopsy or bone marrow specimens from aspiration or biopsy.
- For leukemia only, code 1 when the diagnosis is based only on the complete blood count (CBC), white blood count (WBC) or peripheral blood smear. Do not use code 1 if the diagnosis was based on immunophenotyping or genetic testing using tissue, bone marrow, or blood.
- Code 2 when the microscopic diagnosis is based on cytologic examination of *cells* (rather than tissue) including but not limited to spinal fluid, peritoneal fluid, pleural fluid, urinary sediment, cervical smears and vaginal smears, or from paraffin block specimens from concentrated spinal, pleural, or peritoneal fluid. These methods are rarely used for hematopoietic or lymphoid tumors.
- Code 3 when there is a histology positive for cancer AND positive immunophenotyping and/or positive genetic testing results. Do not use code 3 for neoplasms diagnosed prior to January 1, 2010.

Dx Confirmation - Instructions

57

- Code 5 when the diagnosis of cancer is based on laboratory tests or marker studies which are clinically diagnostic for that specific cancer, but no positive histologic confirmation.
- Code 6 when the diagnosis is based only on the surgeon's report from a surgical exploration or endoscopy or from gross autopsy findings without tissue or cytological findings.
- Code 8 when the case was diagnosed by any clinical method that can not be coded as 6 or 7.
- A number of hematopoietic and lymphoid neoplasms are diagnosed by tests of exclusion where the tests for the disease are equivoical and the physician makes a clinical diagnosis based on the information from the equivoical tests and the patient's clinical presentation.

Disease Progression

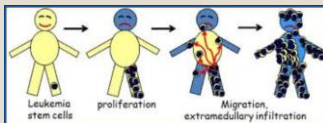
58

- The worsening of a disease over time
- Advancing stage of disease with/out treatment
- Progression from a solitary site of involvement to multiple sites of involvement.
- May be used to describe the progression of a chronic state of disease to an acute state.

Hematopoietic Disease Progression

59

- Same
 - Cell type
 - "Function"
 - Genetics
- Change
 - Symptoms
 - Treatment Approach
 - Prognosis or Life Expectancy



Source: www.haematologica.org

Hematopoietic Disease Progression

60

- Solitary plasmacytoma to plasma cell myeloma
- Smoldering myeloma to plasma cell myeloma
- Early stage/asymptomatic Small Lymphocytic Lymphoma (SLL) or Chronic Lymphocytic Leukemia (CLL) to late stage/symptomatic CLL requiring tx

Transformation

61

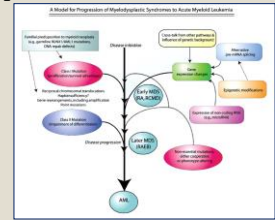
- Change in nature, function, or condition of cells
- Change in cell's potential or type; cell undergoing genetic transformation
- Most transformations are myeloid neoplasms transforming from chronic myeloproliferative or myelodysplastic disease into acute myeloid leukemia
- Chronic Lymphocytic Leukemia (CLL) to Acute Lymphoblastic Leukemia (ALL) is rare - new primary

Hematopoietic Disease Transformation

62

Rare in Lymphoid Neoplasms

- Different
 - Cell type
 - "Function"
 - Genetics
- Change
 - Symptoms
 - Treatment Approach
 - Prognosis or Life Expectancy



Source: www.haematologica.org

Cutaneous Lymphomas

63

- Most primary skin lymphomas are T-cell lymphoma
 - Often multiple skin sites involved - plaque
 - Mycosis Fungoides
 - Sezary Syndrome
- Primary B-cell lymphoma of skin is rare
 - Cutaneous Follicle Center Lymphoma
 - Cutaneous Marginal Zone B-cell lymphoma
 - Cutaneous Diffuse Large B-cell lymphoma
- Diffuse Large B-cell lymphoma of skin is very rare



Tools and Rules



2014 UPDATES 2014

HEMATOPOIETIC DATA BASE

HEME/LYMPH RULES AND INSTRUCTIONS

2014 Data Base Updates 2014

65

- New Format
- New User's Guide
- Content Updates
 - Typos fixed
 - Additional information added
 - MP Calculator Algorithm Updated
 - Information resorted (alphabetical)
 - Transformations Corrected/Enhanced
 - Transformation "to"
 - Transformation "from"
- Enhanced Search Gives Score for Match
- Enhanced Internal Links to Related Rules



2014 Data Base Updates 2014

66

2014 Updates 2014

57

2014 Updates 2014

58

2014 Updates 2014

59

2014 Updates 2014

70

2014 Updates 2014

71

2014 Updates 2014

72

2014 Updates 2014

73

Transformations

None

Transformations From

- 9812.1 Lymphopneumoblastoma (classical Hodgkin's lymphoma)
- 9812.2 Lymphopneumoblastoma (classical Hodgkin's lymphoma)
- 9812.3 Lymphopneumoblastoma (classical Hodgkin's lymphoma)
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None Primary Sites

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None

What's In The Manual/Database

74

Manual (Rules/Instructions)	Database (Dx Yr)
• Introduction	• Neoplasm Name/Definition
• General Instructions	• Alternate Names
• Diagnostic Confirmation	• MP Calculator/Special Rules
• Reportability Instructions	• Primary Site(s)
• Multiple Primary Rules	• Diagnostic Method(s)
• Primary Site Rules	• Abstractor Notes
• Histology Coding Rules	• Immunophenotype
• Grade Coding Rules	• Genetic Tests
• Glossary	• Standard Treatment(s)
• Appendix A - Hx of Coding	• Transformation(s) "to" and "from"
• Appendix B - WHO Lineages	• ICD-O/ICD-9/ICD-10 Codes
• Appendix C - Nodal Chains	• Signs and Symptoms
• Appendix D - Terms / Codes	• Diagnostic Exams
• Appendix E - Obsolete Codes	• Recurrence and Metastases
• Appendix F - Not Reportable	• Epidemiology/Mortality

2014 Rule Updates 2014

75

- Only 1 Format – TEXT
- All Changes to Rules are Effective for Cases Dx 2010>
 - Some Rules Combined
 - Duplicate Rules Removed
 - Corrections to Some Rules
 - Clarifications to Some Rules
 - Example: Review of 2010 and 2011 data shows multiple occurrences of patients with multiple MDS histologies (9980/3, 9982/3, 9983/3, 9984/3, 9985/3, 9986/3, 9989/3, 9991/3, 9992/3)
- Improved (embedded) Navigation to Related Rules
 - Example: See Module 5, PH9 and PH10 for information regarding primary site and histology



2014 Rule Updates 2014

76

- PH Rules Reduced from 43 to 31
- Primary Site Coding Rules for Lymphoma are More Clear
- Lymphoid Combinations Clarified
 - DLBCL with any other lymphoma coded to DLBCL
 - Other mixed lymphomas handled differently
- OBS (obsolete) codes
 - All OBS codes are obsolete as of 1/1/2010
 - OBS codes are now date driven
 - Instruction to use for "DCO's, path only and minimal information" cases removed



2014 Updates 2014

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Hematopoietic and Lymphoid Neoplasm Coding Manual

Effective with Cases Diagnosed 1/1/2010 and Forward

Published January 2014

Editors: Jennifer Ruhl, MD/CA, BHI, CCS, CFR, NCI SEER
Margaret Poggi, MSW, BS, SAS, BHI, CFR, NCI SEER
Lisa Dixon, CFR, NCI SEER
Lore Sore, MD, CFR, NCI SEER

Developer/consultant: Carol Hahn Johnson, BS, CFR, Consultant

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2014 Updates 2014

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Steps to Primary Order for Using the SEER DB and Hematopoietic Coding Manual

Identify the working (primary) histology code(s)

Search the SEER DB using a unique word in the diagnosis, for example "prostate" if the diagnosis is prostate adenocarcinoma (histology code 8590/3), on the computer case (diagnosis). For example, "ovary adenocarcinoma". The results of searched terms that are displayed for each search will be one of the following:

1. The search engine will display every entry with all of the words "ovary" "adenocarcinoma" and "histology". The results displayed ("diagnosis search of ovary") will be all of these words in the histology codes. The results will be reported in one part of the screen (diagnosis, abstractor notes, transformation, etc.).
2. The search engine can also display the number of diagnoses having at least one of the search words by choosing "diagnosis search any term".
3. You can also search on other criteria such as ADL, for some neoplasms (leukemia, DLBCL, for diffuse large B-cell lymphoma, or AML, for acute myeloid leukemia).

When assignments are displayed, click on the desired one to go from information to information to display the record.

Denominator for number of patients using the working histology code(s) with the M code in the manual.

Verify the working histology code(s) using the PH rules in the manual (see Note 1 below).

When the PH rules lead you to a different histology code, open that code in the SEER DB search box and display the record for that histology.

Denominator primary site (see Note 1 below).

The primary site code displayed under Primary Site(s) is the only site code to be used for that histology.

For some neoplasms, only one primary site code is displayed.

1. All leukemia, myeloid leukemias, multiple myeloma and the other myeloid leukemias are assigned primary site have secondary C41. There are an exception. The rule is dependent on ICD-9, as 997.

2. When there is an primary site code listed under Primary Site(s):

- a. Read the Abstractor Notes to find the most common primary site, list unique primary sites, and other sites of assignment for stages II, III, and IV lymphomas. Use the Abstractor Notes to confirm that the site histology combination matches that in the denominator.

Assignment to the manual record is possible. You may also seek a physician's help in determining the primary site.

Note 3. Use Module 10 (PH10) to help determine primary site and histology. Includes: if an histology specific. The message are:

- a. Module 10 AD lymphomas
- b. Module 10 ALL non-Hodgkin lymphomas (NHL) and acute myeloid leukemia (AML)
- c. Module 10 ALL non-Hodgkin lymphomas (NHL) and acute myeloid leukemia (AML)

Denominator for grade. See the Grade Field in the SEER DB.

See the **Grade Field** in the manual when grade cannot be coded using the SEER DB.

Use the Hematopoietic Multiple Primary Calculators in the SEER DB only when instructed by the rules in the Hematopoietic Coding Manual.

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How to Use and Follow the Rules

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Rules Basics

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1. Is the condition reportable?
2. How many cases do I abstract?
3. How do I code the primary site?
4. How do I code the histology?
5. How do I code the grade?



Determining Primary Site

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Primary Site and Histology Coding Rules

1. The primary site and histology coding rules are divided into modules. The first six cover primary site and histology, while the last three cover coding primary site only. Each module covers a group of related hematopoietic or lymphoid neoplasms. However, a specific histology may be covered in more than one module.
2. Go to the first module that fits the case being abstracted. If the situation in the case is not covered in that module, continue on to the next module. Note: The modules are NOT hierarchical, but the rules within each module are in hierarchical order. Apply the rules within each module in order. Stop at the first rule that applies.

Module 1: Post-Transplant Lymphoproliferative Disorder (PTLD)

Post-transplant lymphoproliferative disorder (971.3)

Rule PB1 Code the primary site to the site of origin (lymph node(s) or lymph node region(s), tissue(s), or organ(s)) and code the histology of the accompanying lymphoma or plasmacytoma syndrome when the diagnosis of post-transplant lymphoproliferative disorder and any B-cell lymphoma, T-cell lymphoma, Hodgkin lymphoma, or plasmacytoma syndrome occur simultaneously.

Note 1: These neoplasms are immunologic post-transplant lymphoproliferative disorders. The diagnosis may or may not include the word "immunologic" for post-transplant PTLD, see the definition (971.3).

Note 2: The patient must have a history of a solid organ transplant or an allogeneic bone marrow transplant.

Note 3: Most cases of PTLD occur within a year of transplantation, however, they can occur anytime after the transplant.

Note 4: Monoclonal PTLD is also caused by the immunosuppressive drug. Patients are treated for the lymphoma or plasmacytoma syndrome.

Example: Previous history of kidney transplant. New diagnosis of bone marrow biopsy. BM positive for B-cell lymphoma. Abdominal mass biopsy was positive for PTLD. Immunophenotype and aggressive B-cell malignancy. Immunohistochemistry shows the B-cell malignancy to be Burkitt lymphoma. Code the histology to Burkitt lymphoma and primary site to the abdominal lymph node. (C77.2).

Determining Primary Site

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Module 2: Plasmacytoma (PHT) - PH

Extramedullary plasmacytoma (974.2)
Solitary plasmacytoma of bone (973.1)

Rule PH1 Code the primary site to the site of origin (lymph node(s) or lymph node region(s), tissue(s), or organ(s)) and code the histology extramedullary plasmacytoma (974.2) when any of the following occurs in a site other than bone:

- Extramedullary (extramedullary) plasmacytoma
- Multiple extramedullary (extramedullary) plasmacytoma
- Solitary plasmacytoma
- Plasmacytoma NOS

Note 1: Extramedullary and extramedullary sites include:

- Intestine or other sites including the GI tract, lymph node, bladder, CNS, breast (breast), testis, prostate, and skin.
- The site of origin of CLL, non-multiple myeloma (973.1), or plasmacytoma NOS (974.2).

Note 2: Pathology report a solitary plasmacytoma ranged from 1.4 to 1.6, in terms of number. Code the primary site as site of bone of back (C86) and histology to plasmacytoma (974.2).

Example 1: Bone from plasmacytoma in the vertebral body. Biopsy confirms plasmacytoma. Code the primary site vertebral body (C19) and histology to plasmacytoma (974.2).

Rule PH2 Code the primary site to the specific bone (C80-C89) when the plasmacytoma originated and code the histology solitary plasmacytoma of bone (973.1) when the diagnosis:

- Multiple plasmacytoma
- Multiple plasmacytoma of bone
- Solitary plasmacytoma
- Solitary plasmacytoma of bone

Note 1: Plasma cell myeloma has been removed from the disease codes for the 973.1. See abstractor notes for 973.1 or the non-reportable list, Appendix C10.

Note 2: The exact common site was based with active bone marrow biopsies, in order of frequency they include vertebrae, ribs, skull, pelvis, femur, tibia, and skull.

Note 3: This change from the non-reportable list was implemented in the new ICD-10 code set present in the CDR.

Note 4: Do not code primary site to head (C83), bone marrow (C81), extramedullarized ovum, NOS (C82), or the hematopoietic system, NOS (C84).

Rule PH4 Code the primary site to bone, NOS (C83) and histology solitary plasmacytoma, NOS (973.1) when the only information is that the patient had a plasmacytoma, NOS or solitary plasmacytoma, NOS.

Note 1: When the only information available is that the patient had a plasmacytoma, default to coding plasmacytoma of bone. "Plasmacytoma, NOS" is in a separate list in the New 2013 for 974.2.

Example: Death certificate only note (cause or regional entity only) with underlying cause of death listed as plasmacytoma.

Determining Primary Site

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Module 4: Lymphoma/Leukemia (specific neoplasms that can manifest as either leukemia or lymphoma or both leukemia and lymphoma) PH1 - PH

(972.7, 981.3, 981.5, 981.7, 982.3, 982.7)

Adult T-cell leukemia/lymphoma (HTLV-1 positive) (982.7)

Adult T-cell leukemia/lymphoma (982.7)

B lymphoblastic leukemia/lymphoma with hyperplasia (Hypertrophic CLL) (981.5)

B lymphoblastic leukemia/lymphoma with hyperplasia (Hypertrophic CLL) (981.5)

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Determining Primary Site

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Module 7: Coding Primary Site for Lymphomas Only (PH18 - PH27)

(980.0-972.8, 973.5-974.3, 981.3-981.5, 981.7, 982.3, 982.7)

Note 1: Primary site lymphomas are possible, however, most of the time it is a metastatic site.

Note 2: Do not simply code the site of a biopsy; use the information available from scans to determine the correct primary site.

Rule PH18 Code the primary site to the specified lymph node region when the site of lymphoma is described only as a mass.

Note 1: Metastatic lymph nodes (C77) when the site of the lymphoma is described both as a metastatic mass.

Note 2: Intra-abdominal lymph nodes (C77) when the site of the lymphoma is described only as a retroperitoneal mass or mesenteric mass.

Note 3: Inguinal lymph nodes (C77) when the site of the lymphoma is described only as an inguinal mass.

Note 4: Pelvic lymph nodes (C77) when the site of the lymphoma is described only as a pelvic mass.

Rule PH19 Code the primary site to the specific lymph node region when only one lymph node or one lymph node region is involved.

Note 1: Use this rule when there is bilateral involvement of lymph nodes.

Note 2: See Appendix C for help identifying lymph node names, chains, regions, and codes.

Note 3: Code involvement of intra-abdominal lymph node chain, inguinal lymph node chain, and para-aortic lymph node chain to intra-abdominal lymph node (C77).

Example 1: Code involvement of cervical lymph node chain and mandibular lymph node chain to lymph nodes of head, face and neck (C77).

Example 2: Code involvement of cervical lymph node chain to lymph nodes of head, face and neck (C77).

Rule PH21 Code the primary site to multiple lymph node regions, NOS (C78) when multiple lymph node regions are defined by ICD-O-3, are involved and it is not possible to identify the lymph node region where the lymphoma originated.

Note 1: See Table B12.1 when there is also organ involvement.

Note 2: Do not simply code the site of a biopsy; use the information available from scans to determine the correct primary site. See Primary Site Coding Appendix C for help identifying lymph node names, chains, regions, and codes.

Note 3: See Appendix C for help identifying lymph node names, chains, regions, and codes.

Example 1: Cervical (C77) and mandibular (C77) lymph nodes involved with B-cell lymphoma. Code the primary site to lymph nodes of multiple regions (C78).

Example 2: CT scan showed involvement of the cervical lymph nodes (C77) and the mandibular lymph nodes (C77). No additional involvement was identified during the work-up. Biopsy of a cervical lymph node confirmed lymphoma. Code the primary site to lymph nodes of multiple regions (C78).

Rule PH22 Code the primary site to lymph nodes, NOS (C77) when:

• Lymphoma is present in an organ and lymph nodes that are not reported for that organ and the organ of the lymphoma cannot be determined even after consulting the physician OR

• Lymphoma is present in more than one organ and the regional nodes for all organs involved OR

Single Node Station/Multiple LN/Extranodal

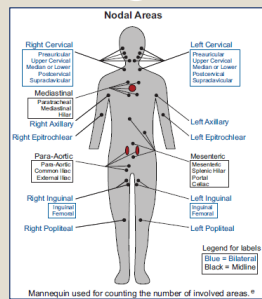
85

- Biopsy Site
- Single Node Station
- Bilateral - Same Node Station?
- Multiple Node Stations
- No nodal involvement



Number of Involved Nodal Areas

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Source: NCCN.org and Dana-Farber Cancer Institute, Inc.

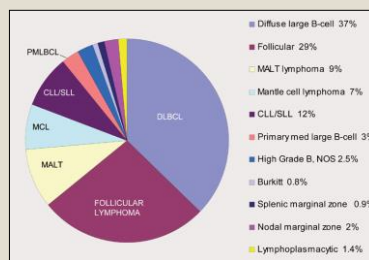
Determining Histologic Type

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- Code the non-specific (NOS) histology when – PH28
- Code the specific histology when – PH29
- Use the Heme Data Base in Most Cases – PH30
- Code the Numerically Higher – PH31

B-Cell Lymphoid Histology Distribution in Adults

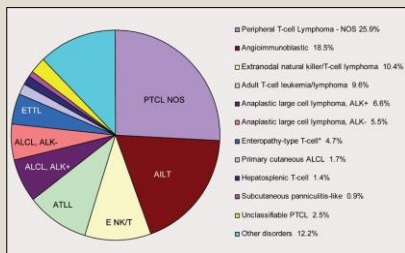
88



Source: WHO Classification of Hematopoietic and Lymphoid Neoplasms

T-Cell Lymphoid Histology Distribution in Adults

89



Source: WHO Classification of Hematopoietic and Lymphoid Neoplasms

Determining Grade/Differentiation

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Grade of Tumor Marker

There is a [table](#) of [algorithms](#) for coding Grade, Differentiation, or Cell Subtype (NACCR/NCI Sites # 44) that is to be implemented for cases diagnosed January 1, 2014. Students often need an update on the Immunohistochemistry Code rules below.

Prerequisite List for Coding Grade or Differentiation

This is a prerequisite for with Site 1 having the **Subject prefix**.

Site 1: Immunohistochemistry (IHC) done on the specimen and result for the site provided in the Heme report.

Site 2: Immunohistochemistry (IHC) done on the specimen and result for the site provided in the Heme report.

Note: IHC done on Site 3 on pages 1451 of ICD-O-3, Immuno grade. This table is updated.

Note 3: The physician comment is code phenotype on the grade field, use extension three, one part of medical record including but not limited to:

- Pathology report
- Genetic analysis
- Case notes
- Final diagnosis
- Flow data

Note 4: There have been physician comment, code Grade/Phenotype 3 Columns.

Note 5: The site, right-click under the Immunohistochemistry site, 1, 2, 3, 4, 5, 6, 7, 8.

Note 6: If the site is not complete, the site, immunohistochemistry, is a grade code in the Tumor Grade field. This table also in the Working Foundation category of ImmuHem Diagnosis. Do not code 1, 2 or 3 including Molecular ImmuHem.

Rule 01 - Code cell type and Assessment, use coded, use appropriate, code 0 for the following specific/differentiation assignment, use 9 for Indeterminate/unknown, 0 for benign, unspecified/differentiation, histologic, and ductal cell assignment.

9100-9109: Solitary metastases of site
 9110-9119: Systemic metastases
 9120-9129: Solid cell metastases
 9130-9139: Carcinoid tumor metastases
 9140-9149: Sarcoma
 9150-9159: Embryonal, embryonic cell, carcinoma
 9160-9169: Embryonal, embryonic cell, sarcoma
 9170-9179: Embryonal, embryonic cell, carcinoma
 9180-9189: Embryonal, embryonic cell, sarcoma
 9190-9199: Acute myeloid leukemia
 9200-9209: Acute myeloid leukemia
 9210-9219: Acute myeloid leukemia
 9220-9229: Acute myeloid leukemia
 9230-9239: Acute myeloid leukemia
 9240-9249: Acute myeloid leukemia
 9250-9259: Acute myeloid leukemia
 9260-9269: Acute myeloid leukemia
 9270-9279: Acute myeloid leukemia
 9280-9289: Acute myeloid leukemia
 9290-9299: Acute myeloid leukemia
 9300-9309: Acute myeloid leukemia
 9310-9319: Acute myeloid leukemia
 9320-9329: Acute myeloid leukemia
 9330-9339: Acute myeloid leukemia
 9340-9349: Acute myeloid leukemia
 9350-9359: Acute myeloid leukemia
 9360-9369: Acute myeloid leukemia
 9370-9379: Acute myeloid leukemia
 9380-9389: Acute myeloid leukemia
 9390-9399: Acute myeloid leukemia

Hematopoietic and Lymphoid Neoplasm Coding Manual 40

Appendices

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- Appendix A - History of Hematopoietic /Lymphoid Coding
- Appendix B - WHO Classification - Lineage Tables
- Appendix C - Lymph Node/Lymph Node Chain Table
- Appendix D - New Histology Terms and Codes
- Appendix E – Obsolete Hematopoietic Codes
- Appendix F – Non-Reportable Terms - NEW

Training

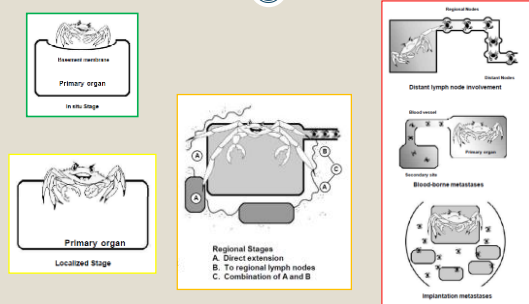
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NEW Hematopoietic and Lymphoid Neoplasm Training
<https://educate.fhcr.org>



Solid Tumor Staging

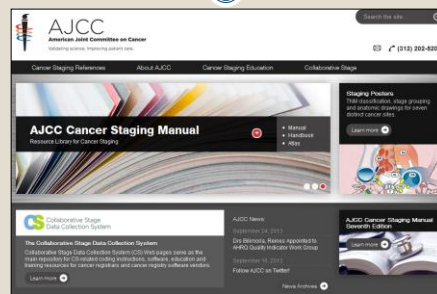
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Source: SEER Summary Staging Manual 2000

AJCC Cancer Staging - TNM

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<http://www.cancerstaging.org>

AJCC Cancer Staging - TNM

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<http://www.cancerstaging.org>

CS COLLABORATIVE STAGE DATA COLLECTION SYSTEM

CS Schemas for Lymphoid Neoplasms:

- Heme/Retic
- Lymphoma
- Lymphoma Ocular Adnexa
- Mycosis Fungoides
- Myeloma Plasma Cell Disorder

<http://www.cancerstaging.org/cstage/index.html>

CSv02.04 Cancer Schema

Announcements

- CSv02.04 Release Announcement
- CSv02.04 Support for Known Issues
- Documentation SSOs for v02.04

Educational Resources

- TS Ext. Eval. 1 or 3 for Op. Findings
- TS Ext. Eval. 18 and SSX. 18
- TS Ext. Eval. 18 and SSX. 18

<http://www.cancerstaging.org/cstage/index.html>

ORDER

- Alphabetical Order
- Natural Order

Site	Site	Site
Lip, Lipop.	Melanoma Pharynx Oropharynx	Stomach
Melanoma Lip Lower	Melanoma Pharynx Oropharynx	Testis
Lip Lower	GST Pancreatic	Melanoma Penis
Melanoma Lip Lower	Squamous GI Larynx	Melanoma Penis Other
Lip Other	Stomach	Penis
Melanoma Lip Other	GI Stomach	Micro Cell Penis
Trachea, Bron.	NET Stomach	Prostate
Melanoma Tongue Base	Small Intestine	Testis
Truncus Arterio.	GI Small Intestine	Melanoma Testis
Melanoma Tongue Arterio.	NET Small Intestine	Melanoma Testis Subepididymic
Uterus Uteri.	Appendix	Melanoma Testis Subepididymic
Melanoma Uterus Upper	Carotid Artery	Larynx Other
Uterus Lower	Colon	Melanoma Uterus Other
Melanoma Uterus Lower	GI Colon	Uterus
Uterus Other	GI Appendix	Bladder
Melanoma Vag Other	GI Colon	Urinary Other
Vag Mouth	NET Colon	Urinary Other
Melanoma Vag Mouth	Rectum	Uterus Cervix
Vagina Vulva	GI Rectum	Melanoma Uterine Cervix
Melanoma Vulva Vulva	NET Rectum	Eye Other
	None	Melanoma Eye
		Melanoma Eye Study

<http://www.cancerstaging.org/cstage/index.html>

HemeRetic Schema

Hematopoietic, Reticuloendothelial, Immunoproliferative, and Myeloproliferative Neoplasms

- M - See list of specific histologies below. All primary sites (C00.0-C00.9) are included unless otherwise specified.
- Schema includes only preferred terms from ICD-O-3.
- Plasmacytomas (9731 and 9734) and Multiple Myeloma (9732), except for cases with primary site C441, C690 and C695-C696, have been moved to the Myeloma/PlasmaCellDisorder schema in V0203.
- 9733 Plasma cell leukemia [except C441, C690, C695-C696]
- 9740 Mest cell sarcoma
- 9741 Malignant histiocytosis
- 9742 Mest cell leukemia
- 9746 Malignant histiocytosis
- 9752 Langerhans cell histiocytosis, "unifocal" (see new reportable code 9751:3)
- 9753 Langerhans cell histiocytosis, "multifocal" (see new reportable code 9751:3)
- 9754 Langerhans cell histiocytosis disseminated
- 9755 Histiocytic sarcoma
- 9756 Langerhans cell sarcoma
- 9757 Interdigitating dendritic cell sarcoma
- 9758 Follicular dendritic cell sarcoma
- 9760 Immunoproliferative disease, NOS
- 9761 Waldenstrom macroglobulinemia
- 9762 Heavy chain disease, NOS
- 9764 Immunoproliferative small intestine disease
- 9765 Monoclonal gammopathy of undetermined significance*
- 9768 Amyloidosis, immunoproliferative lesion*
- 9767 Angioimmunoblastic lymphadenopathy*
- 9768 T gamma immunoproliferative disease*
- 9769 Immunoglobulin deposition disease*
- 9800 Leukemia, NOS
- 9801 Acute leukemia, NOS
- 9805 Acute lymphocytic leukemia
- 9809 Lymphoid leukemia, NOS [except C441, C690, C695-C696]
- 9823 B cell chronic lymphocytic leukemia/small lymphocytic lymphoma [C420, C421, or C424 ONLY]
- 9826 Burkitt cell leukemia/lymphoma [except C441, C690, C695-C696]
- 9827 Adult T cell leukemia/lymphoma (HTLV-1 positive) [C420, C421, or C424 ONLY]
- 9832 Polychromic leukemia, NOS [except C441, C690, C695-C696]
- 9833 Polychromic leukemia, B cell type [except C441, C690, C695-C696]

<http://www.cancerstaging.org/cstage/index.html>

HemeRetic Schema

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Code	Description
100	Localized disease (Single/solitary/unifocal/isolated): May be coded for: Mast cell sarcoma (9740) Malignant histiocytosis (9750) Langerhans cell histiocytosis (9751) Histiocytic sarcoma (9755) Langerhans cell sarcoma (9756) Dendritic cell sarcoma (9757, 9758) Myeloid sarcoma (9930)
800	Systemic disease (All histologies including those in 100)
999	Unknown, extension not stated Primary tumor cannot be assessed Not documented in patient record

<http://www.cancerstaging.org/cstage/index.html>

Lymphoma Staging

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Diaphragm

stage I stage II stage III stage IV

Source: <http://cancer.gov>

Lymphoma Staging

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Stage	Description
*Reprinted with permission from AJCC: <i>Hodgkin and non-Hodgkin lymphomas</i> . In: <i>Edge SB, Byrd DR, Compton CC, et al., eds. AJCC Cancer Staging Manual</i> . 7th ed. New York, NY: Springer, 2009, pp 607-62. [§2]	
I	Involvement of a single lymphatic site (i.e., nodal region, Waldeyer's ring, thymus, or spleen) (I), or localized involvement of a single extralymphatic organ or site in the absence of any lymph node involvement (IE).
II	Involvement of two or more lymph node regions on the same side of the diaphragm (II), or localized involvement of a single extralymphatic organ or site in association with regional lymph node involvement with or without involvement of other lymph node regions on the same side of the diaphragm (IIIE).
III	Involvement of lymph node regions on both sides of the diaphragm (III), which also may be accompanied by extralymphatic extension in association with adjacent lymph node involvement (IIIE) or by involvement of the spleen (IIIIS) or both (IIIE.S).
IV	Diffuse or disseminated involvement of one or more extralymphatic organs, with or without associated lymph node involvement; or isolated extralymphatic organ involvement in the absence of adjacent regional lymph node involvement, but in conjunction with disease in distant sites). Stage IV includes any involvement of the liver or bone marrow, lungs (other than by direct extension from another site), or cerebrospinal fluid.
Designations applicable to any stage	
A	No symptoms.
B	Fever (temperature ≥38°C), drenching night sweats, unexplained loss of ≥10% of body weight within the preceding 6 months.
E	Involvement of a single extranodal site that is contiguous or pretentious to the known nodal site.
S	Spleen involvement.

Source: <http://cancer.gov>

Lymphoma Staging

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Source: AJCC Cancer Staging Atlas, 2nd edition

Lymphoma Staging

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Source: AJCC Cancer Staging Atlas, 2nd edition

Lymphoma Staging

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Source: AJCC Cancer Staging Atlas, 2nd edition

Lymphoma Staging

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Source: AJCC Cancer Staging Atlas, 2nd edition

Lymphoma Schema

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Collaborative Stage for TNM 7 - Revised 10/25/2011

Lymphoma
Hodgkin and Non-Hodgkin Lymphomas of All Sites (excluding Mucositis Fungoides and Sezary Disease)

- M 9590-9595, 9702-9729, 9735-9737, 9739 (EXCEPT C44.1, C49.0, C59.5-C59.6)
- M 9811-9816, 9820, 9827, 9837 (EXCEPT C42.0, C42.1, C42.4, C44.1, C59.0, C59.5-C59.6)

<ul style="list-style-type: none"> CS_Tumor_Size = 000 CS_Extension CS_Tumor_Size/Ext_Eval CS_Lymph_Nodes CS_Lymph_Nodes_Ext = 0 CS_Regional_Nodes_Positive = 00 CS_Regional_Nodes_Extensive = 00 CS_Mets_MDI = 0 CS_Mets_Ext = 0 CS_Site-Specific_Factor 1 Associated with HIV/AIDS CS_Site-Specific_Factor 2 Systemic_Symptoms_at_Diagnosis CS_Site-Specific_Factor 3 International_Prognostic_Index_(IPI) CS_Site-Specific_Factor 4 Follicular_Lymphoma_Prognostic_Index_(FLIPI) CS_Site-Specific_Factor 5 International_Prognostic_Score_(IPS) 	<ul style="list-style-type: none"> CS_Site-Specific_Factor 7 = 000 CS_Site-Specific_Factor 8 = 000 CS_Site-Specific_Factor 9 = 000 CS_Site-Specific_Factor 10 = 000 CS_Site-Specific_Factor 11 = 000 CS_Site-Specific_Factor 12 = 000 CS_Site-Specific_Factor 13 = 000 CS_Site-Specific_Factor 14 = 000 CS_Site-Specific_Factor 15 = 000 CS_Site-Specific_Factor 16 = 000 CS_Site-Specific_Factor 17 = 000 CS_Site-Specific_Factor 18 = 000 CS_Site-Specific_Factor 19 = 000 CS_Site-Specific_Factor 20 = 000 CS_Site-Specific_Factor 21 = 000 CS_Site-Specific_Factor 22 = 000 CS_Site-Specific_Factor 23 = 000 CS_Site-Specific_Factor 24 = 000 CS_Site-Specific_Factor 25 = 000
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Source: <http://cancerstaging.org>

Lymphoma Schema

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100	involvement of a single lymph node region Stated as Stage I
110	Localized involvement of a single extralymphatic organ/site in the absence of any lymph node involvement Multifocal involvement of one extralymphatic organ/site Stated as Stage IE
120	Involvement of spleen only Stated as Stage IS
200	Involvement of two or more lymph node regions on the SAME side of the diaphragm Stated as Stage II
210	Localized involvement of a single extralymphatic organ/site WITH involvement of its regional lymph node(s) WITH or WITHOUT involvement of other lymph node(s) on the SAME side of the diaphragm Direct extension to adjacent organs or tissues Stated as Stage IIE
220	Involvement of spleen PLUS lymph node(s) BELOW the diaphragm Stated as Stage IIS

<http://www.cancerstaging.org/cstage/index.html>

Plasma Cell Neoplasm Staging

109

Table 1: The Durie-Salmon Staging System for Multiple Myeloma

Stage	Hemoglobin	Calcium	Myeloma Protein	Bone Lesions
I ^a	>10 g/dL	Normal or ≤12 g/dL	IgG peak <5 g/dL IgA peak <3 g/dL Bence-Jones protein <4 g/24 h	None or solitary bone plasmacytoma only
II ^b	Not I or III	Not I or III	Not I or III	Not I or III
III ^c	<8.5 g/dL	>12 mg/dL	IgG peak >7 g/dL IgA peak >3 g/dL Bence-Jones protein >12 g/24 h	>3 lytic lesions

^a Stage I must demonstrate all of the criteria.
^b Stage II defined as all patients who do not qualify as Stage I or III.
^c Stage III must demonstrate one or more of the criteria.
 Source: Reference 7.

MyelomaPlasmaCellDisorder Schema

110

Collaborative Stage for TMM 7 - Revised 10/25/2011

MyelomaPlasmaCellDisorder

Plasma Cell Disorders including Myeloma

- 9731 Plasmacytoma, NOS (except C441, C500, C505, C506)
- 9732 Multiple myeloma (except C441, C500, C505, C506)
- 9734 Plasmacytoma, extramedullary (except C441, C500, C505, C506)
- Note 1: This schema was added in V2003. Originally these histologies were part of the Hemofitic schema.
- Note 2: AJCC does not define TMM staging for this site.

CS Tumor Stage = 900	CS Site Specific Factor 7 = 900
CS Extension	CS Site Specific Factor 8 = 900
CS Tumor Site/Ext Eval = 9	CS Site Specific Factor 9 = 900
CS Lymph Nodes	CS Site Specific Factor 10 = 900
CS Lymph Nodes Eval = 9	CS Site Specific Factor 11 = 900
Regional Nodes Positive = 99	CS Site Specific Factor 12 = 900
Regional Nodes Examined = 99	CS Site Specific Factor 13 = 900
CS Metn at US	CS Site Specific Factor 14 = 900
CS Metn Eval = 9	CS Site Specific Factor 15 = 900
CS Site Specific Factor 1	CS Site Specific Factor 16 = 900
OBsolete - Janus Kinase 2 (JAK2) (also known as JAK2 Exon 12)	CS Site Specific Factor 17 = 900
CS Site Specific Factor 2	CS Site Specific Factor 18 = 900
Durie-Salmon Staging System	CS Site Specific Factor 19 = 900
CS Site Specific Factor 3	CS Site Specific Factor 20 = 900
Multiple Myeloma Terminology	CS Site Specific Factor 21 = 900
CS Site Specific Factor 4 = 900	CS Site Specific Factor 22 = 900
CS Site Specific Factor 5 = 900	CS Site Specific Factor 23 = 900
CS Site Specific Factor 6 = 900	CS Site Specific Factor 24 = 900

<http://www.cancerstaging.org/cstage/index.html>

MyelomaPlasmaCellDisorder Schema

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- Note 1: Osseous plasmacytomas are localized tumors occurring in the bone. There may be soft tissue extension.
- Note 2: Extrasosseous (extramedullary) plasmacytomas are plasma cell neoplasms that arise in tissues other than bone. The most common sites are the upper respiratory tract, the gastrointestinal tract, lymph nodes, bladder, central nervous system (CNS), breast, thyroid, testis and skin.

<http://www.cancerstaging.org/cstage/index.html>

MyelomaPlasmaCellDisorder Schema

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- Note 3: Criteria for the diagnosis of multiple myeloma include: presence of clonal bone marrow plasma cells or plasmacytoma, presence of an M-protein in serum and/or urine, and the presence of related organ or tissue impairment. Do not use this criteria to determine the diagnosis of multiple myeloma. Code according to histologic confirmation or physician statement according to the AJCC 7th edition.
- Note 4: Multiple myeloma or plasma cell myeloma is a widely disseminated plasma cell neoplasm, characterized by a single clone of plasma cells derived from B cells that grows in the bone marrow. It is always coded to 810 or 820 for systemic involvement.

<http://www.cancerstaging.org/cstage/index.html>

MyelomaPlasmaCellDisorder Schema

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Code	Description
100	OBsolete DATA RETAINED V2003 Localized disease (except solitary/ambicubic/biastromatocytic), may be coded for: Plasmacytoma, NOS (9731) (solitary myeloma) Plasmacytoma, extramedullary (9734) (not occurring in bone)
110	Single plasmacytoma lesion WITHOUT soft tissue extension or unknown if soft tissue extension (9731)
200	Single plasmacytoma lesion WITH soft tissue extension (9731)
300	Single plasmacytoma lesion occurring in tissue other than bone (9734)
400	Multiple osseous or multiple extrasosseous plasmacytoma lesions (9731, 9734)
500	Plasmacytoma, NOS (9731) Not stated if osseous or extrasosseous
600	OBsolete DATA RETAINED V2003 Systemic disease (diffuse-infiltrative) All histologies including those in 100
810	Plasma cell myeloma/multiple myeloma/hypermucos (9732)
820	Myeloma, NOS Excludes plasma cell myeloma or multiple myeloma (see code 810)
	Unknown: extension not stated Primary tumor cannot be assessed Not documented in patient record

Annotations: A green vertical bar highlights codes 110, 200, 300, and 400. An orange arrow points to code 9734. A red arrow points to code 9732. A red 'X' is placed over the bottom row.

Site Specific Factors - Lymphoma

114

- SSF1 – Associated with HIV/AIDS
- SSF2 – Systemic Symptoms at Diagnosis
- SSF3 – International Prognostic Index (IPI)
- SSF4 – Follicular Lymphoma Prognostic Index (FLIPI)
- SSF5 – International Prognostic Score (IPS)

Site Specific Factors – Plasma Cell Tumors

115

- SSF1 – OBSOLETE
- SSF2 – Durie-Salmon Staging System
- SSF3 – Multiple Myeloma Terminology

000	Multiple myeloma/Plasma cell myeloma with no other modifiers Multiple myeloma, NCS, Myeloma, NCS
010	Asymptomatic myeloma
020	Early or evolving myeloma
030	Inactive, indolent, or smoldering myeloma
080	Other terminology describing myeloma
100	Any combination of terms in codes 010-080

Treatment Options – Lymphoid Neoplasms

116

- Hodgkin Lymphoma
- Non-Hodgkin Lymphoma
- Chronic Lymphocytic Leukemia
- Acute Lymphocytic Leukemia
- Other Lymphoid Neoplasm



Source: Mosaic Rainbow and Woodland Forest - <http://www.etsy.com>

Treatment Options – Basic Concepts

117

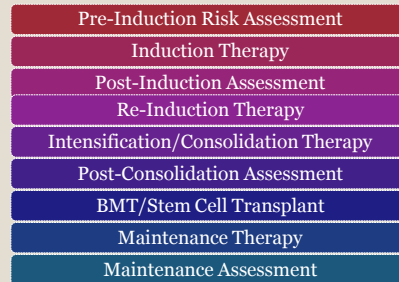
- Surgery
- Chemotherapy
- Radiation Therapy
- Hormonal Therapy
- Combination Therapy
- Continuation Therapy
- Bone Marrow/Stem Cell Transplant



Image Source: <http://greenplanetparadise.com> and <http://avinomilerner.com>

Treatment Options – Basic Concepts

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Treatment Options – Basic Concepts

119

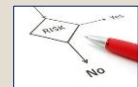


Source: <http://cancer.gov> – Pediatric Myeloid Neoplasm NCI PDQ for Health Professionals

Treatment Options – Basic Concepts

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- Risk-Based Treatment – Pre-Induction Risk
 - Patient Characteristics
 - ✦ Performance Status
 - ✦ Age at Diagnosis
 - ✦ Comorbidities
 - ✦ B-Symptoms
 - Neoplasm Characteristics
 - ✦ Morphology
 - ✦ Immunophenotype
 - ✦ Stage of Differentiation
 - ✦ Molecular/Cyto-Genetics
 - Special Characteristics of Neoplasm or Patient

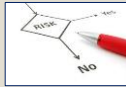


Source: <http://cancer.gov> – Pediatric Lymphoid Neoplasm NCI PDQ for Health Professionals

Treatment Options – Basic Concepts

121

- Risk-Based Treatment – Induction Failure
 - Identify patients at highest risk of induction failure:
 - T-cell phenotype (especially without a mediastinal mass)
 - B-precursor ALL with very high presenting leukocyte counts
 - Bulky Disease
- Risk-Based Treatment – Re-Induction/Consolidation
 - Re-Induction
 - Intensification
 - Consolidation
- Risk-Based Treatment – Sanctuary Sites
- Risk-Based Treatment – Maintenance Therapy



Source: <http://cancer.gov> – Pediatric Lymphoid Neoplasm NCI PDQ for Health Professionals

Treatment Options – Basic Concepts

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- Risk-Based Treatment Assessment Examples
 - Low Risk Disease – Stage I, II – no B symptoms, no bulky disease
 - Intermediate Risk Disease – Stage I, II with B symptoms
 - Intermediate Risk Disease – Stage I, II with bulky disease
 - Intermediate Risk Disease – Stage IIIA, IVA
 - High Risk Disease – Stage IIIB, IVB
 - High Risk Disease – Poor response to initial chemotherapy

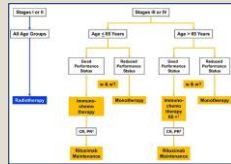


Source: <http://cancer.gov> – Pediatric Lymphoid Neoplasm NCI PDQ for Health Professionals

Treatment - Surgery

123

- Surgery – when do you code for lymphoma?
- Surgery – when do you NOT code for lymphoma?
- Why the difference?
- When is Surgery = TX
- Why so seldom?



Treatment - Chemotherapy

124

- Chemotherapy Regimens
- REMINDER: Many regimens contain Prednisone which is to be coded under Hormone Therapy – in addition to the combination Chemotherapy



Treatment - Chemotherapy

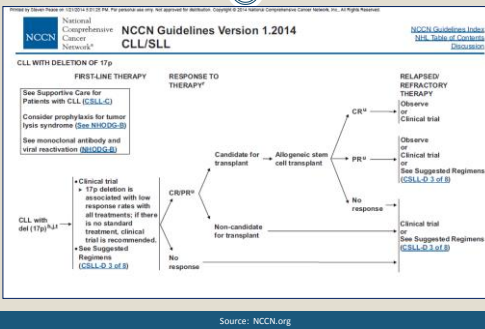
125

Treatment Options – CLL/SLL

126

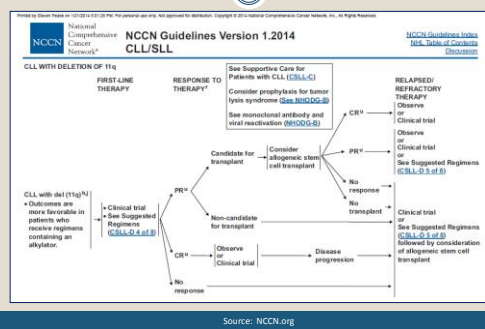
Treatment Options – CLL/SLL (del 17p)

127



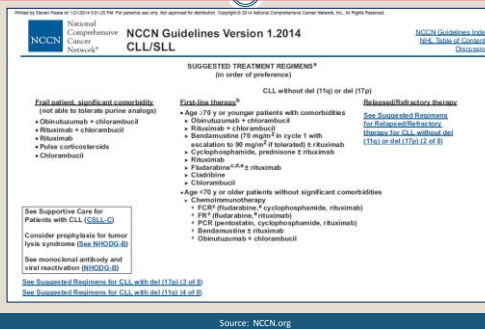
Treatment Options – Lymphoma

128



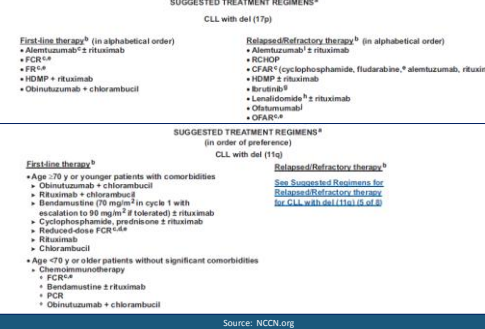
Treatment Options – Lymphoma

129



Treatment Options – Lymphoma

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FDA Approved Agents for HL / NHL (A-L)

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Abitrexate (Methotrexate)	Cyclophosphamide
Adcetris (Brexitinib Vedotin)	Cytosar (Cyclophosphamide)
Adriamycin PFS (Doxorubicin Hydrochloride)	Denileukin Difitox
Adriamycin RDF (Doxorubicin Hydrochloride)	DepoCyt (Liposomal Cytarabine)
Ambochlorin (Chlorambucil)	Doxorubicin Hydrochloride
Ambochlorin (Chlorambucil)	DTIC-Dome (Dacarbazine)
Arranon (Nelarabine)	Folex (Methotrexate)
Bendamustine Hydrochloride	Folex PFS (Methotrexate)
Bexcar (Tositumomab and Iodine I 131 Tositumomab)	Folotyn (Pralatrexate)
Blenoxane (Bleomycin)	Ibritumomab Tuxetan
Bleomycin	Ibrutinib
Bortezomib	Imbruvica (Ibrutinib)
Brentuximab Vedotin	Intron A (Recombinant Interferon Alfa-2b)
Chlorambucil	Intodax (Romidepsin)
Clafen (Cyclophosphamide)	Lenalidomide

Source: www.cancer.gov/cancertopics/druginfo

FDA Approved Agents for HL / NHL (M-Z)

132

Leukan (Chlorambucil)	Revlimid (Lenalidomide)
Linfolin (Chlorambucil)	Rituxan (Rituximab)
Liposomal Cytarabine	Rituximab
Matulane (Procarbazine Hydrochloride)	Romidepsin
Methotrexate	Tositumomab and Iodine I 131 Tositumomab
Methotrexate LPF (Methotrexate)	Trenda (Bendamustine Hydrochloride)
Mexate (Methotrexate)	Velban (Vincristine Sulfate)
Mexate-AQ (Methotrexate)	Velcade (Bortezomib)
Mozobil (Plerixafor)	Velcar (Vincristine Sulfate)
Nelarabine	Vinblastine Sulfate
Neszar (Cyclophosphamide)	Vincasar PFS (Vincristine Sulfate)
Ontak (Denileukin Difitox)	Vincristine Sulfate
Plerixafor	Vorinostat
Pralatrexate	Zevalin (Ibritumomab Tuxetan)
Recombinant Interferon Alfa-2b	Zolinza (Vorinostat)

Source: www.cancer.gov/cancertopics/druginfo

Common Chemo Regimens in NHL

Regimen	Regimen
CHOP	EPOCH
COPP	ICE
CVP	R-CHOP

C	Cyclophosphamide
H	Doxorubicin Hydrochloride
O	Vincristine Sulfate (Oncovin)
P	Prednisone
P	Procarbazine Hydrochloride
V	Vincristine Sulfate (Oncovin)
E	Etoposide
I	Ifosfamide
C	Carboplatin
R	Rituximab

Source: www.cancer.gov/cancertopics/druginfo

Chemo Regimens in Hodgkin Lymphoma

Regimen	Regimen
ABVD	ICE
ABVE	MOPP
ABVE-PC	OEPA
BEACOPP	OPPA
COPP	Stanford V
COPP-ABV	VAMP

A	Adriamycin
B	Bleomycin
V	Vinblastine Sulfate
D	Dacarbazine
E	Etoposide
P	Prednisone
C	Cyclophosphamide
V	Vincristine Sulfate (Oncovin)
P	Procarbazine Hydrochloride
I	Ifosfamide
C	Carboplatin
M	Methotrexate

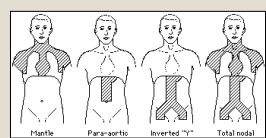
Source: www.cancer.gov/cancertopics/druginfo

Treatment - BRM

- Biological Response Modifiers – when and why?
- SEER*Rx is Primary Reference
- Examples:
 - Rituximab – cytostatic monoclonal antibody – CLL, NHL
 - Belinostat – histone deacetylation inhibitor – CLL, MM, NHL
 - Thalidomide – antiangiogenic agent – MM, leukemia
 - Epratuzumab – NOT BRM – Radioisotope – Code RT – NHL
 - Zevalin – NOT BRM – Radiolabeled monoclonal antibody – NHL

Treatment - Radiation

- Radiation Therapy – when and why?



Treatment Strategy

```

    graph TD
      IL[Indolent Lymphoma] --> S3[Stage III]
      IL --> S4[Stage IIIIV, Relapsed or Progressive Disease]
      S3 --> RT[Radiation Therapy]
      S3 --> AS[Asymptomatic]
      S3 --> S4
      AS --> WW[Watch and Wait or Chemotherapy]
      S4 --> ST[Single or Combination Therapy]
            
```

Treatment - Radiation

PRINCIPLES OF RADIATION THERAPY*

- Field:**
 - Treatment with photons, electrons, or protons may all be appropriate, depending upon clinical circumstances.
 - Isotopic-irradiation therapy (ISIT):** See table above.
 - ISRT is recommended as the appropriate field for NHL. Planning for ISRT requires modern CT-based simulation and planning capabilities, incorporating either modern imaging like PET + MRI with either enhanced field delineation, incorporating either modern imaging like PET + MRI with either enhanced field delineation.
 - ISRT targets the site of the originally involved lymph node(s). The field encompasses the original suspicious volume prior to chemotherapy or surgery. Yet, it spares adjacent uninvolved organs (like lung, bone, muscle, or kidney) when lymphadenopathy regresses following chemotherapy.
 - The pre-chemotherapy or pre-surgery gross tumor volume (GTV) provides the basis for determining the clinical target volume (CTV). Concerns for quantitative volume of disease and uncertainties in original imaging accuracy or localization may lead to expansion of the CTV and are determined individually using clinical judgment. Possible movement of the target by respiration as determined by 4D-CT or functional (stereotaxic) target volume (TV) should also influence the field CTV.
 - The planning treatment volume (PTV) is an additional expansion of the CTV that accounts only for setup variations (see KRJ definitions).
 - Organ at risk (OAR) should be outlined for optimizing treatment plan decisions.
 - The treatment plan is designed using conventional, 3-D conformal, or IMRT techniques using clinical treatment planning considerations of coverage and dose reductions for OAR.
- ISRT for extracranial disease:**
 - Similar principles as for ISRT nodal sites (see above).
 - For most organs and particularly for indolent disease, the whole organ alone is the CTV (eg, stomach, salivary gland, orbit, thyroid, breast, testis).
 - For nonindolent, localized sites, only the involved part of the organ is irradiated with adequate margins.
 - For most NHL, subacute or radiation is required for uninvolved lymph nodes.

General Dose Guidelines	Considerations after chemotherapy or RT
• Localized CLL/ALL: 24-36 Gy	• Diffuse large cell lymphoma or PTCL: 30-36 Gy
• Follicular lymphoma: 24-30 Gy	• Complications after chemotherapy: CR: 30-36 Gy
• Marginal zone lymphoma: 30 Gy	• Complementary after PR: 40-50 Gy
• Diffuse: 30 Gy	• RT as primary treatment for indolent or non-metastatic disease for chemotherapy: 45-55 Gy
• Other extranodal sites: 24-30 Gy	• Salvage pre- or post-relapse cell transplantation: 30-40 Gy
• Nodal MCL: 24-30 Gy	• Primary cutaneous anaplastic large cell lymphoma: 30-36 Gy
• Early stage mantle cell lymphoma: 30-36 Gy	
• Minimum RT 45 Gy if 2 mos be required for palliative/local control of SLL, FL, MZL, MCL	

Treatment - Radiation

NCCN Guidelines Version 1.2014 Follicular Lymphoma (grade 1-2)

STAGE	INITIAL THERAPY	RESPONSE TO THERAPY	RECOMMENDATIONS
Stage I, II	BRT ¹ (preferred for clinical stage I or contiguous stage II)	CR ¹ or PR ¹	See monoclinal antibody and viral reactivation (NCCN 1-2)
		NR	See Stage I, II, III, IV, V (FOLL-1)
	Immunotherapy ± chemotherapy (See FOLL-1)	CR ¹ or PR ¹	See Stage I, II, III, IV, V (FOLL-2)
		NR	See Stage I, II, III, IV, V (FOLL-2)
Observation (selected cases) ³	CR ¹ or PR ¹	See Stage I, II, III, IV, V (FOLL-2)	
	NR	See Stage I, II, III, IV, V (FOLL-2)	

¹H&P and labs every 3-6 mo for 5 y and then annually or as clinically indicated surveillance imaging².
²Up to 2 y post completion of treatment; CT scans no more than every 6 mo. ³+2 y, no more than annually.
⁴Progressive disease:^{1,4} see Stage I, II, III, IV, V (FOLL-1). For transformation, see FOLL-1.

Treatment - Other

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- Other Therapy – when and why?
- PUVA for cutaneous lymphoma



Text Documentation

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DATA ITEMS REQUIRING COMPLETE TEXT DOCUMENTATION	
Date of DX	RX Summ – Surg Prim Site
Seq No	RX Summ – Scope Reg LN Surgery
Sex	RX Summ – Surg Oth Reg/Distant
Primary Site	RX Date – Surgery
Subsite	RX Summ – Radiation
Laterality	Rad Rx Modality
Histologic Type	RX Date – Radiation
Behavior Code	RX Summ – Chemo
Grade	RX Date – Chemo
	RX Summ – Hormone
CS Tumor Size	RX Date – Hormone
CS Ext	RX Summ – BRM/Immunotherapy
CS Tumor Ext/Eval	RX Date – BRM/Immunotherapy
Regional Nodes Positive	RX Summ – Transplant/Endocrine
Regional Nodes Examined	RX Date – Transplant/Endocrine
CS LN	RX Summ – Other
CS LN Eval	RX Date - Other
CS Mets	
CS Mets Eval	Any Unusual Case Characteristics
All FCDS Req'd SSEs	Any Pertinent Patient/Family History

References

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- **2014 Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual**, J. Ruhl, M. Adamo, C. Hahn Johnson, L. Dickie, NCI SEER, 2014
- **Classification, Characteristics, and Behavior of Myeloid Neoplasms**, G.M. Dores, NCI, 2010
- **WHO Classification of Tumors of Hematopoietic and Lymphoid Tissues**, 4th ed, S. Swerdlow, E. Campo, N. Lee Harris, E. Jaffe, S. Pileri, H. Stein, J. Thiele, J. Vardiman, IARC, Lyon, FR, 2008
- **National Comprehensive Cancer Network (NCCN) 2014 Clinical Practice Guidelines – NHL, ALL, Myeloma, and Hodgkin Lymphoma**
- **The 2008 WHO Classification of Lymphoid Neoplasms and Beyond**; E. Campo, S. Swerdlow, NL Harris, E Jaffe; Blood 2011 117
- **A Revised European-American Classification of Lymphoid Neoplasms**; NL Harris, E Jaffe, H Stein; Blood 1994 84
- **FCDS Data Acquisition Manual**

Questions

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