Abstracting and Coding Lymphoid Neoplasms

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

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?

- 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
Pediatric Neoplasms

Adult Neoplasms

WHO Definition

- "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

- **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

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Lymphoid Neoplasm Characteristics

- **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

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Common Lymph Node Chains for Lymphoma

Source: CancerHelpUK.org
Extra-Nodal Lymphoma

Common Types of Lymphoma

Causes and Risk Factors

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

Causes and Risk Factors

Gene Mutation in Familial ALL

- 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

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

“B” Symptoms

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

Immune System

- 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
Immune System

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

Hematopoiesis

- 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

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

Lymphoid Cell Line Differentiation

- 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
The Lymphatic System

Lymphatic Organs

- 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

http://commonsensehealth.com
Classification of Lymphoid Neoplasms

- 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

- 1982 – Working Formulation
- 1994 – Revised European-American Classification of Lymphoid Neoplasms
2008 WHO Classification of Lymphoid Neoplasms

Incorporates:
- Histology/Morphology
- Stage of Differentiation
- Immunophenotype
- Genotypic features
- Clinical features

2008 WHO Classification - Lymphoid

Precursor Lymphoid Neoplasm

Mature B-Cell Neoplasm

Mature B-Cell Neoplasm (con't)

Mature T-Cell and NK-Cell Neoplasm
2008 WHO Classification - Lymphoid

- Hodgkin Lymphoma
- Histiocytic/Dendritic Cell Neoplasm
- PTLD (Post-Transplant)

Understanding Complex Disease Processes

- Lymph Node Biopsy
- Extranodal Site Biopsy
- Diagnostic Imaging (CT/PET/MRI)
- Bone Marrow Aspirate
- Bone Marrow Biopsy
- Histology/Morphology
- Immunohistocytohemistry
- Flow Cytometry (Immunophenotype)
- Cytogenetics
- Molecular Genetic Studies
  - FISH
  - PCR

The Clinical Workup

- 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

Table 2
Diagnostic criteria for plasma cell myeloma

<table>
<thead>
<tr>
<th>Symptomatic plasma cell myeloma</th>
<th>Asymptomatic (smoldering) myeloma</th>
</tr>
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<tbody>
<tr>
<td>M-protein in serum or urine*</td>
<td>M-protein in serum at myeloma levels (&gt; 30 g/L) and/or ≥ 10% clonal plasma cells in BM</td>
</tr>
<tr>
<td>BM clonal plasma cells or plasmacytoma†</td>
<td>No related organ or tissue impairment end-organ damage or bone lesions (CRAB) or myeloma-related symptoms</td>
</tr>
<tr>
<td>Related organ or tissue impairment heavy chain disease: (CRAB)</td>
<td></td>
</tr>
</tbody>
</table>

Source: BLOOD, 12 MAY 2011 VOLUME 117, NUMBER 19

Plasma Cell Neoplasms

Immunophenotype

- 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

Cluster of Differentiation Markers – B Cell

Cluster of Differentiation Markers – T Cell
Dx Confirmation - Codes

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Positive biopsy 1</td>
</tr>
<tr>
<td>2</td>
<td>Positive biopsy 2</td>
</tr>
<tr>
<td>3</td>
<td>Positive biopsy 3</td>
</tr>
</tbody>
</table>

Dx Confirmation - Instructions

1. There is no prior necessity for coding Diagnostic Confirmation for hematopoietic and lymphoid tissues. However, for specific histologic types diagnosis is confirmed by immunophenotyping or genetic testing. See the Neoplasms Databases (US) for information on the definitive diagnostic confirmation for specific types of tumors.

2. Code 1 when the aspirate diagnosis is based on tissue specimens from biopsy, fine needle, or other methods of tissue biopsy. Specimens from aspiration or biopsies.

3. For histologic 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, fine needle aspirate, or biopsied tissue.

4. Code 2 when the aspirate diagnosis is based on cytopathic examination of cell or tissue (other than tissue) excluding not limited to: gram stain, gram-stained film, gram-stained smear, or cervical smear and vaginal smear; or from paraffin block specimens from sonographically guided or other smear or sentinel nodes. These methods are used for hematopoietic or lymphoid tumors.

5. Code 3 when there is a histological positive for cancer and/or positive immunophenotyping and/or positive genetic testing results. Do not use code 3 for tumors diagnosed prior to January 1, 2014.

6. Code 4 when the diagnosis of cancer is based on laboratory tests or other methods which are clinically diagnostic for that specific cancer, but lack positive histologic confirmation.

7. Code 5 when the diagnosis is based only on the surgeon’s report from a surgical exploration or malignancy or from gross or autopsied findings without bone marrow biopsy.

8. Code 6 when the cancer is diagnosed by any clinical method that cannot be coded by 1 or 3. A diagnosis of hematopoietic and lymphoid malignancies are diagnosed by tests of exclusion where the tumor for the disease are organized and the patient needs a clinical diagnostic based on the information plus the excluded test and the patient’s clinical presentation.
**Disease Progression**

- 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.

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**Hematopoietic Disease Progression**

**Same**
- Cell type
- “Function”
- Genetics

**Change**
- Symptoms
- Treatment Approach
- Prognosis or Life Expectancy

Source: www.haematologica.org

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**Hematopoietic Disease Progression**

- 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
- 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
- Rare in Lymphoid Neoplasms
- Different
  - Cell type
  - “Function”
  - Genetics
- Change
  - Symptoms
  - Treatment Approach
  - Prognosis or Life Expectancy

Source: www.haematologica.org

Cutaneous Lymphomas
- 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 Update 2014
HEMATOPOIETIC DATA BASE
HEME/LYMPH RULES AND INSTRUCTIONS

2014 Data Base Updates 2014

- 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 Updates 2014

What’s In The Manual/Database

<table>
<thead>
<tr>
<th>Manual (Rules/Instructions)</th>
<th>Database (Dx Yr)</th>
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<tbody>
<tr>
<td>Introduction</td>
<td>Neoplasm Name/Definition</td>
</tr>
<tr>
<td>General Instructions</td>
<td>Alternate Names</td>
</tr>
<tr>
<td>Diagnostic Confirmation</td>
<td>MP Calculator/Special Rules</td>
</tr>
<tr>
<td>Reportability Instructions</td>
<td>Primary Site(s)</td>
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<tr>
<td>Multiple Primary Rules</td>
<td>Diagnostic Method(s)</td>
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<tr>
<td>Primary Site Rules</td>
<td>Abstrator Notes</td>
</tr>
<tr>
<td>Histology Coding Rules</td>
<td>Immunophenotype</td>
</tr>
<tr>
<td>Grade Coding Rules</td>
<td>Genetic Tests</td>
</tr>
<tr>
<td>Glossary</td>
<td>Standard Treatment(s)</td>
</tr>
<tr>
<td>Appendix A – Hx of Coding</td>
<td>Transformation(s)/&quot;to&quot; and &quot;from&quot;</td>
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<tr>
<td>Appendix B – WHO Lineages</td>
<td>ICD-0/ICD-9/ICD-10 Codes</td>
</tr>
<tr>
<td>Appendix C – Nodal Chains</td>
<td>Signs and Symptoms</td>
</tr>
<tr>
<td>Appendix D – Terms / Codes</td>
<td>Diagnostic Exams</td>
</tr>
<tr>
<td>Appendix E – Obsolete Codes</td>
<td>Recurrence and Metastases</td>
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<tr>
<td>Appendix F – Not Reportable</td>
<td>Epidemiology/Mortality</td>
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</table>

2014 Rule Updates 2014

- 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

- 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

Hematopoietic and Lymphoid Neoplasms Coding Manual
Effective with Case Approved CIC Code and Forward

2014 Updates 2014
How to Use and Follow the Rules

Rules Basics

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

Primary Site and Reporting Coding Rules

1. The primary site and histology coding rules are divided into two sections. The first one covers the primary site and histology, while the second one covers other coding basics. Each module covers a group of related requirements or content. However, a specialty (e.g., surgery) may be covered in a single section.
2. The first step is to determine if the case can be abstracted. If the case occurs and is reported in the module, continue up to the next module.
3. Not all modules are NOT standardized, so the case rules must be manual. Apply the rules relevant to the module. Any of the rules can be used.

Module 3: Poorly Differentiated Lymphomas and Leukemias

For example, lymphoma or leukemia (L&L) may be reported as either diffuse or nodular. The diagnostic categories for lymphomas include: DLBCL, nodular sclerosis; follicular lymphoma; mantle cell lymphoma; and anaplastic large cell lymphoma (ALCL).

Note: The primary site in the following example may be ambiguous. However, if the site is specified, it follows the rule. A primary site of the bone should be considered as unspecified. If the bone is not specified, it should follow the rule.
Single Node Station/Multiple LN/Extranodal

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

Number of Involved Nodal Areas

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

Determining Histologic Type

- 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

- Diffuse large B-cell 37%
- Follicular 29%
- MALT lymphoma 9%
- Marginal zone lymphoma 7%
- CLL/SLL 13%
- Primary mediastinal large B-cell 3%
- High Grade B, NOS 2.5%
- Burkitt 0.8%
- Splenic marginal zone 0.6%
- nodal marginal zone 3%
- Lymphoplasmacytic 1.4%

Source: WHO Classification of Hematopoietic and Lymphoid Neoplasms

T-Cell Lymphoid Histology Distribution in Adults

- Peripheral T-cell lymphoma: NOS 25.9%
- Angioimmunoblastic 16.8%
- Cutaneous T-cell lymphoma 19.6%
- Adult T-cell leukemia/lymphoma 9.6%
- Anaplastic large cell lymphoma, ALCL 6.9%
- Anaplastic large cell lymphoma, ALK- 3.8%
- Enteropathy-type T-cell 4.7%
- Primary cutaneous ALC 1.7%
- Hepatosplenic T-cell 1.4%
- Intraepithelial T-cell 0.5%
- Unspecified T-cell, 2.3%
- Other variants 12.1%

Source: WHO Classification of Hematopoietic and Lymphoid Neoplasms

Determining Grade/Differentiation

...
Appendices

- 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

NEW Hematopoietic and Lymphoid Neoplasm Training
https://educate.fhcrc.org

Solid Tumor Staging

Source: SEER Summary Staging Manual 2020
HemeRetic Schema
HemeRetic Schema

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>100</td>
<td>Localized disease: (Single bone/marrow/mucosal isolated) May be coded for: Blast cell leukemia (9749) Malignant lymphomas (9740) Langerhans cell histiocytosis (9715) Histocytic sarcomas (9711) Multiple myeloma (9790) Dendritic cell sarcoma (9735) Myeloid sarcoma (9990)</td>
</tr>
<tr>
<td>800</td>
<td>Systemic disease (All histologies including those in 100)</td>
</tr>
<tr>
<td>999</td>
<td>Unknown, extension not dated Primary tumor cannot be assessed Not documented in patient record</td>
</tr>
</tbody>
</table>


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Lymphoma Staging

![Diagram of Lymphoma Staging](http://cancer.gov)

Source: [http://cancer.gov](http://cancer.gov)

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Lymphoma Staging

![Diagram of Lymphoma Staging](http://cancer.gov)

Source: [http://cancer.gov](http://cancer.gov)
Plasma Cell Neoplasm Staging

<table>
<thead>
<tr>
<th>Stage</th>
<th>Hemoglobin</th>
<th>Calculated</th>
<th>Myeloma Protein</th>
<th>Bone Lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>&gt;15 g/dL</td>
<td>Normal or 812 g/dL</td>
<td>IgG peaks &lt;3 g/dL</td>
<td>None or solitary bone plasmacytomas only</td>
</tr>
<tr>
<td>1</td>
<td>&gt;15 g/dL</td>
<td>812-100 g/dL</td>
<td>IgG peaks &gt;3 g/dL</td>
<td>1 or 2 lesions</td>
</tr>
<tr>
<td>2</td>
<td>&gt;15 g/dL</td>
<td>&gt;100 g/dL</td>
<td>IgG peaks &gt;3 g/dL</td>
<td>1 or 2 lesions, bone involvement</td>
</tr>
<tr>
<td>3</td>
<td>&lt;15 g/dL</td>
<td>&lt;812 g/dL</td>
<td>IgG peaks &gt;3 g/dL</td>
<td>3 or more lesions, bone involvement</td>
</tr>
</tbody>
</table>

Note 1: Osseous plasmacytomas are localized tumors occurring in the bone. There may be soft tissue extension.

Note 2: Extraosseous (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.

MyelomaPlasmaCellDisorder Schema

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

Site Specific Factors - Lymphoma

- 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

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

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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<tbody>
<tr>
<td>M1</td>
<td>Multiple myeloma, no evidence of other myeloma, multiple myeloma, MG, monoclonal, MG</td>
</tr>
<tr>
<td>M2</td>
<td>Asymptomatic myeloma</td>
</tr>
<tr>
<td>M3</td>
<td>Early symptomatic myeloma</td>
</tr>
<tr>
<td>M3E</td>
<td>Excessive or smoldering myeloma</td>
</tr>
<tr>
<td>M3O</td>
<td>Other terminal, non-smoldering myeloma</td>
</tr>
<tr>
<td>M3P</td>
<td>Any combination of terms in codes M1, M2, M3</td>
</tr>
</tbody>
</table>

Treatment Options – Lymphoid Neoplasms

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


Treatment Options – Basic Concepts

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

Treatment Options – Basic Concepts

- Pre-Induction Risk Assessment
- Induction Therapy
- Post-Induction Assessment
- Re-Induction Therapy
- Intensification/Consolidation Therapy
- Post-Consolidation Assessment
- BMT/Stem Cell Transplant
- Maintenance Therapy
- Maintenance Assessment

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 Myeloid Neoplasm NCI PDQ for Health Professionals
Treatment Options – Basic Concepts

- 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](http://cancer.gov)

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Treatment Options – Basic Concepts

- 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 IIA, IVA
  - High Risk Disease – Stage IIIB, IVA
  - High Risk Disease – Poor response to initial chemotherapy

Source:  [http://cancer.gov – Pediatric Lymphoid Neoplasm NCI PDQ for Health Professionals](http://cancer.gov)

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Treatment - Surgery

- 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?

Source:  [http://cancer.gov – Pediatric Lymphoid Neoplasm NCI PDQ for Health Professionals](http://cancer.gov)
Treatment - Chemotherapy

- Chemotherapy Regimens

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

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

<table>
<thead>
<tr>
<th>Agent Name</th>
<th>Drug Class</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leukeran (Chlorambucil)</td>
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<td>Rituxan (Rituximab)</td>
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<td>Vinca Alkaloids</td>
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<td>Blenoxane (Bleomycin)</td>
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<td>Mexican (Mexican)</td>
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<td>Nelarabine (Neosar)</td>
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<td>Folex (Methotrexate)</td>
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<tr>
<td>Velcade (Bortezomib)</td>
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<td>Velban (Vinblastine Sulfate)</td>
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<td>Zolinza (Vorinostat)</td>
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Source: [NCCN.org](https://www.nccn.org)

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

<table>
<thead>
<tr>
<th>Agent Name</th>
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<td>Mozobil (Plerixafor)</td>
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Source: [www.cancer.gov/cancertopics/druginfo](https://www.cancer.gov/cancertopics/druginfo)
Common Chemo Regimens in NHL

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Source: www.cancer.gov/cancertopics/druginfo

CHemo Regimens in Hodgkin Lymphoma

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<td>COPP</td>
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<td>COPP-ABV</td>
<td>VAMP</td>
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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 – Radiolabeled monoclonal antibody – NHL
  - Zevalin – NOT BRM – Radiolabeled monoclonal antibody – NHL
Treatment - Other

- Other Therapy – when and why?

- PUVA for cutaneous lymphoma

Text Documentation

<table>
<thead>
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References

- Classification, Characteristics, and Behavior of Myeloid Neoplasms, G.M. Dores, NCI, 2010
- 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