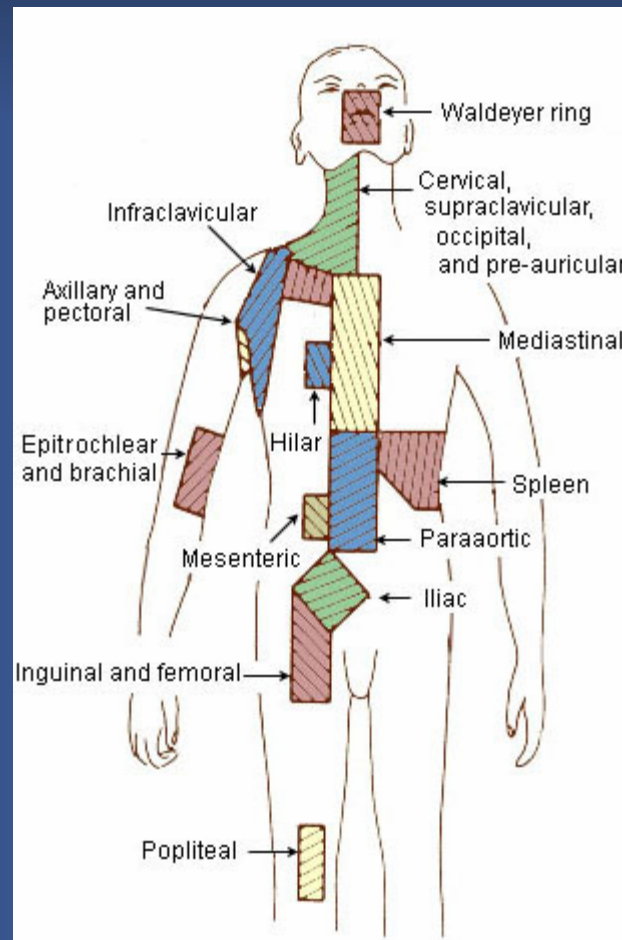


# **NAACCR Central Registry Webinar Series**

**Shannon Vann, CTR  
Jim Hofferkamp, CTR**



# Lymphoma Cancer Surveillance Data Collection



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Image source: SEER Training Website

# Background

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# Bone Marrow

- Present in the soft inner part of some bones such as the skull, shoulder blade, ribs, pelvis, and backbones.
- The bone marrow is made up of blood-forming stem cells, lymphoid tissue, fat cells, and supporting tissues that aid the growth of blood-forming cells.

# Hematopoietic Malignancies

- Lymphoma is a general term for hematopoietic solid malignancies of the *lymphoid* series.
- Leukemia is a general term for liquid malignancies of either the *lymphoid* or the *myeloid* series.

# Lymphoma General Terms

- **Pluripotential hematopoietic stem cells (PHSC)**
  - ◆ The precursor cells which give rise to all the blood cell types of both the myeloid and lymphoid lineages.

# General Terms

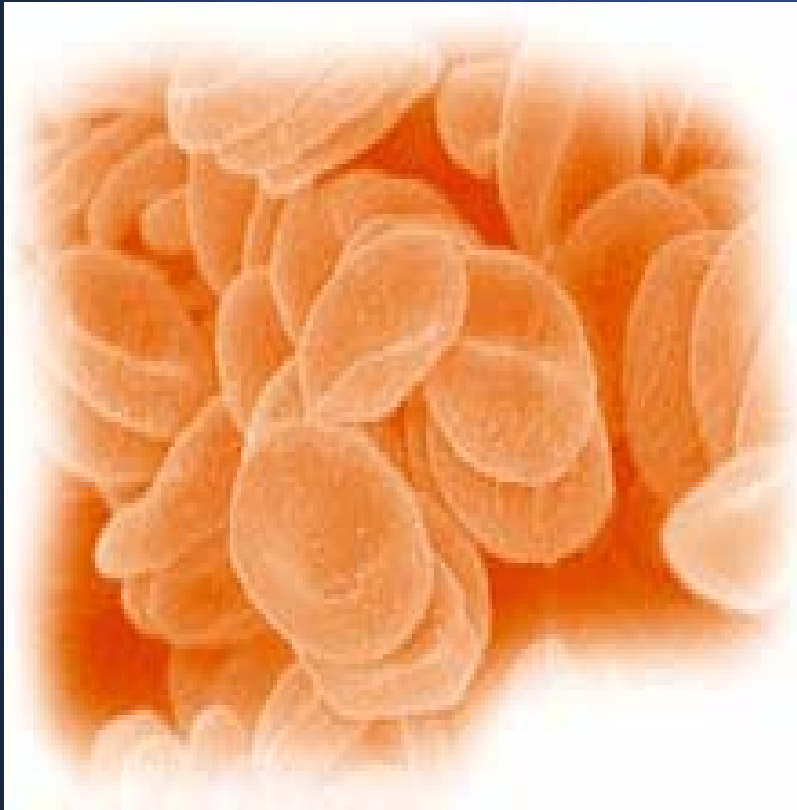
- **Myeloid**

- ◆ These cells are precursors for red blood cells or erythrocytes.

- **Lymphoid**

- ◆ These are the precursors for lymphocytes (a type of white blood cell).

# General Terms



- There are two broad categories of lymphocytes.
  - ◆ T-Cells
  - ◆ B-Cells

Image source: SEER Training Website

# T-Cells and B-Cells

- Immature lymphocytes that travel to the thymus differentiate into T-Cells
  - ◆ "T" is for thymus
- Immature lymphocytes that travel to the spleen or lymph nodes differentiate into B cells
  - ◆ "B" stands for the bursa of Fabricius, which is an organ unique to birds, where B cells mature.

# General Terms

- **Flow Cytometry** can be used to detect antigens on the surface of lymphocytes that can be used to differentiate sub-types.

# General Terms

- These antigens are known as CD molecules (or Clusters of Differentiation).
  - ◆ CD4 and CD8 are characteristic of subtypes of T-Cells
  - ◆ CD20 are characteristic of B-Cells.
  - ◆ The presence of CD56 and/or CD16 and the absence of CD3 indicate an NK (Natural Killer) Cell.

# Anatomy and Physiology

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# Lymphatic System

- **Functions of lymphatic system**
  - ◆ **Return interstitial fluid to blood**
  - ◆ **Absorb fats and vitamins**
  - ◆ **Defend against disease and infection**

# Lymphatic Components

## ■ Lymph

- ◆ Is interstitial fluid similar to and derived from plasma
- ◆ Maintains blood volume and pressure
- ◆ Drains from organs to regional lymph nodes and then to distant lymph nodes

# Lymphatic Components

- **Lymphatic capillaries**
  - ◆ Small vessels that carry fluids away from tissues
- **Lymphatic trunks**
  - ◆ Right lymphatic duct drains upper right quadrant of body
  - ◆ Thoracic duct drains the rest of body

# Lymphatic Components

- **Lymph nodes**
  - ◆ Shaped like small beans
  - ◆ Surrounded by capsule
  - ◆ Divided into nodules containing lymphocytes and macrophages

**Lymph Node Structure**

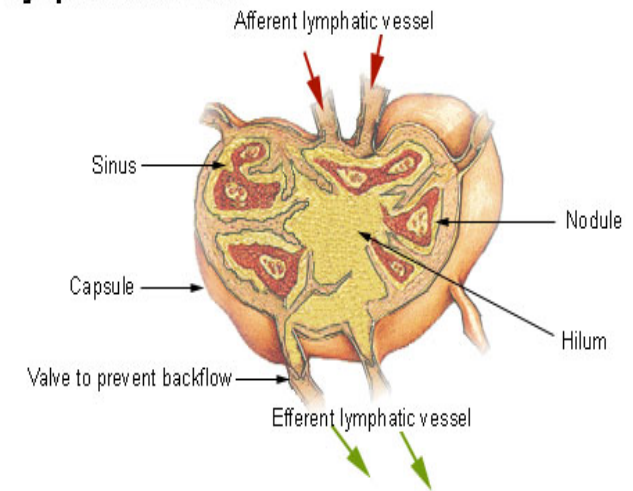
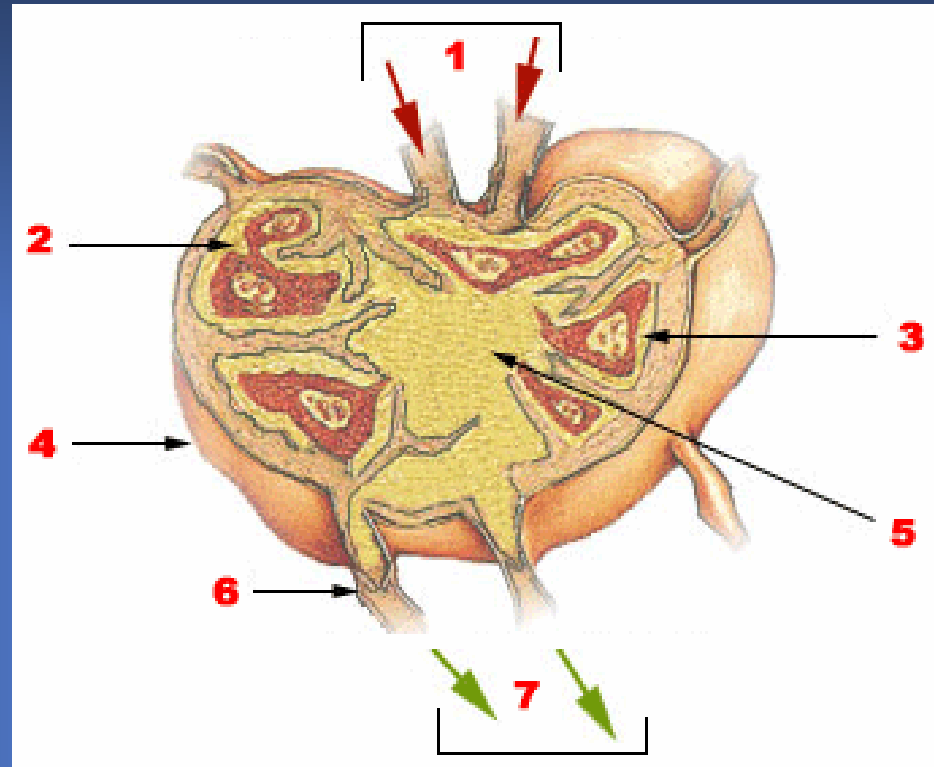


Image source: SEER Training Website

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# Structure of the Lymph Node

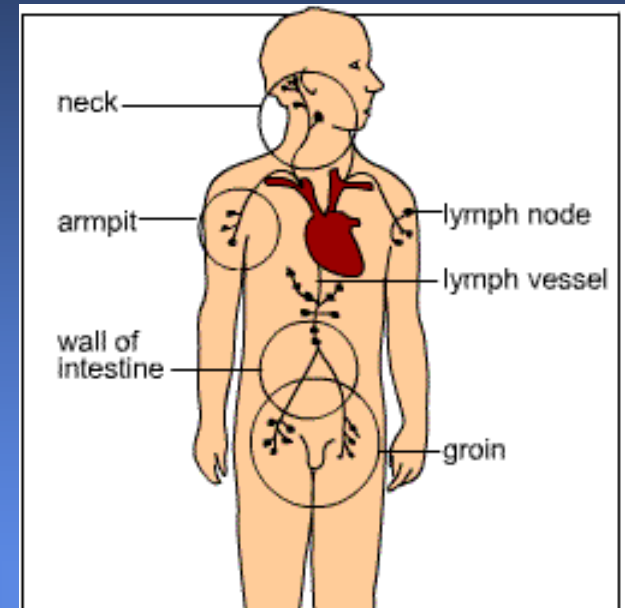
- ◆ 1. Afferent lymphatic vessel
- ◆ 2. Sinus
- ◆ 3. Nodule
- ◆ 4. Capsule
- ◆ 5. Medulla
- ◆ 6. Valve to prevent backflow
- ◆ 7. Efferent lymphatic vessel



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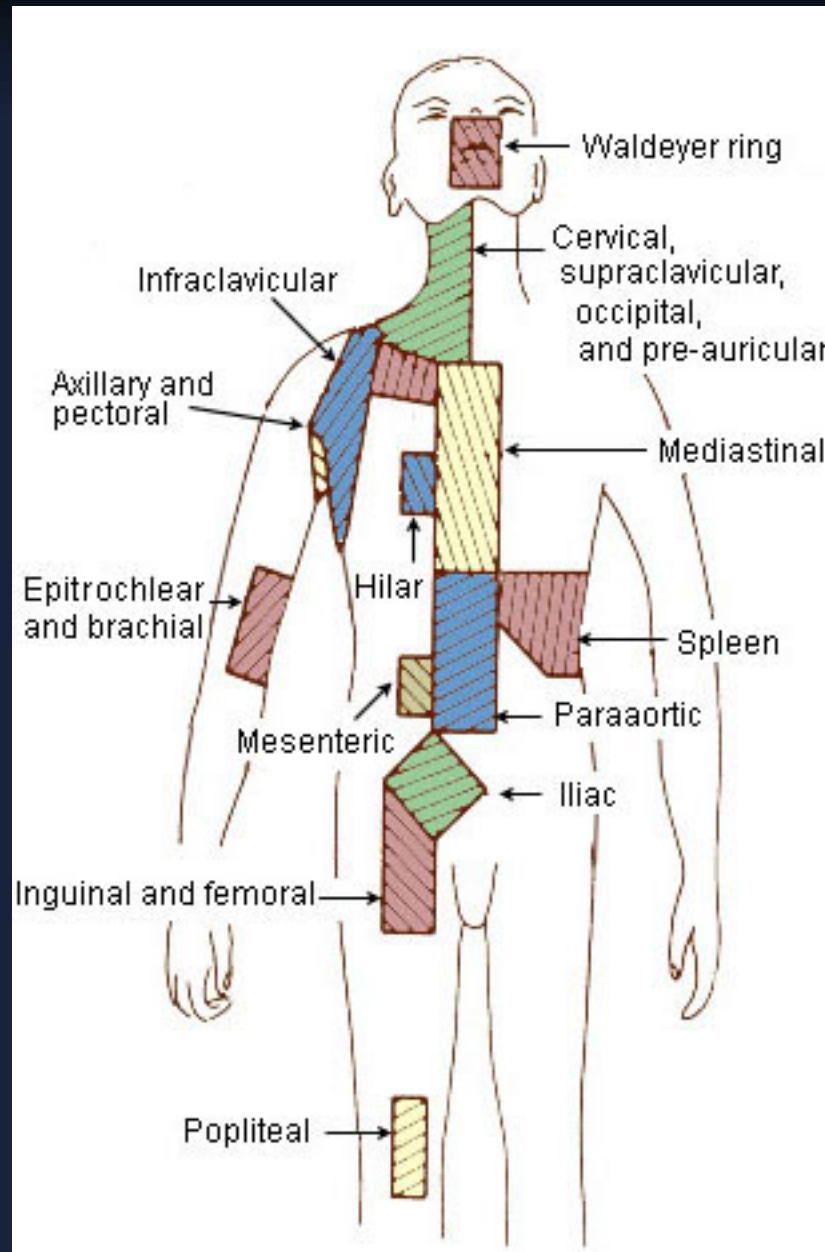
# Lymphatic Components

- Lymphatic organs
  - ◆ Lymph nodes
    - ◆ Armpit: axillary nodes
    - ◆ Neck: cervical nodes
    - ◆ Groin: inguinal nodes



# Lymph Node Regions

- Grouped in and named for body areas
- Parallel to and named for major blood vessels

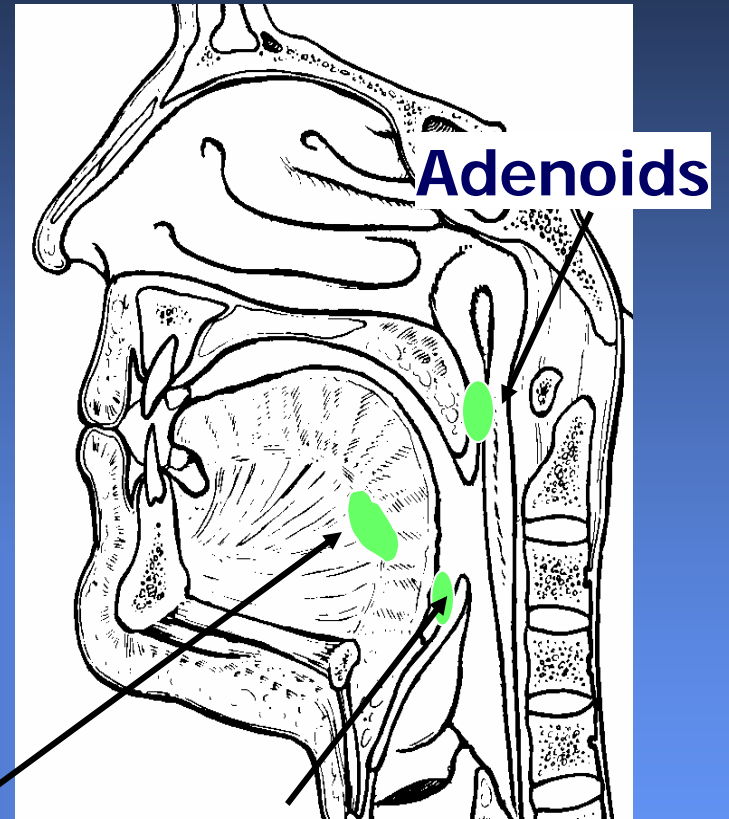


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# Lymphatic Structures

## ■ Tonsils

- ◆ Pharyngeal tonsils (adenoids) C11.1
- ◆ Palatine tonsils C09.9
- ◆ Lingual tonsils C02.4
- ◆ Waldeyer ring C14.2



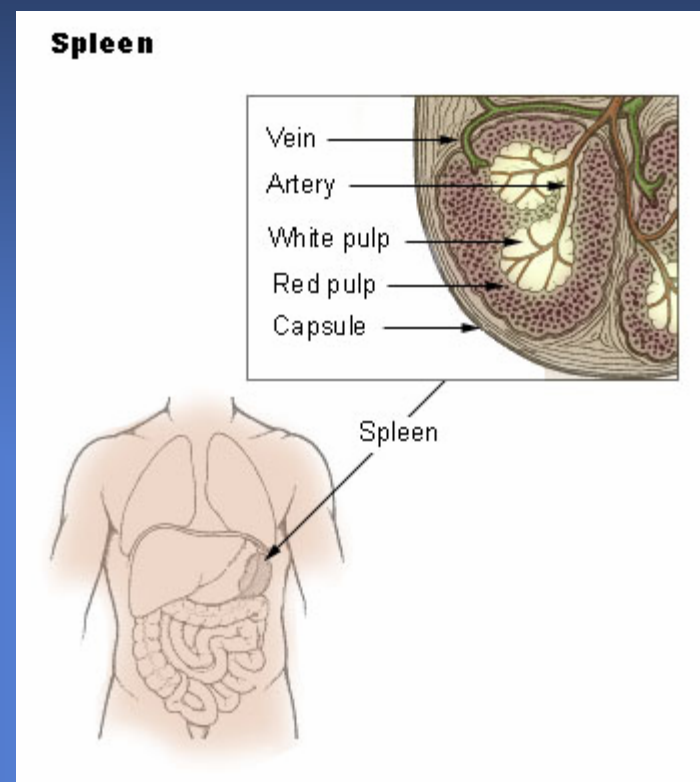
Lingual tonsils Palatine tonsils

# Lymphatic Components

## ■ Lymphatic organs

### ◆ Spleen

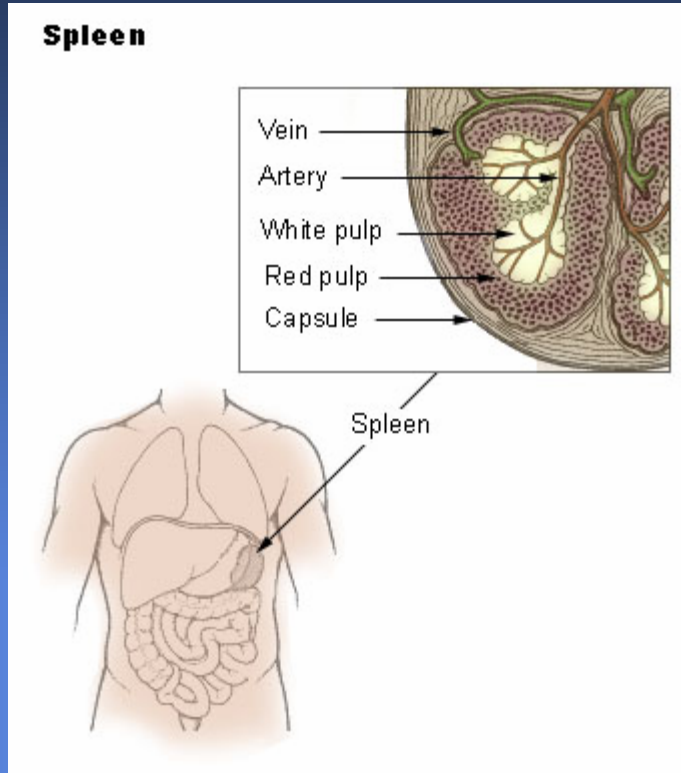
- ◆ Is the largest lymphatic organ
- ◆ Filters blood
- ◆ Contains two major tissues
  - White pulp
  - Red pulp



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# Lymphatic Components

- Lymphatic organs
  - ◆ Thymus
    - ◆ Processes T-cells to fight disease in other body organs
    - ◆ Produces thymosin



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# Lymphatic Components

- Lymphatic organs
  - ◆ Peyer's patches
    - ◆ Lymphoid nodules of the small intestine, primarily the ileum
  - ◆ Lymphoid nodules of the appendix

# Lymphoma and Acquired Immunodeficiency Syndrome (AIDS)

- AIDS defining lymphoma
  - ◆ Burkitt's lymphoma
  - ◆ Immunoblastic lymphoma
  - ◆ Primary central nervous system (CNS) lymphoma
- AIDS related lymphoma
  - ◆ Hodgkin lymphoma
  - ◆ Non-Hodgkin lymphoma

# ICD O 3 Coding

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# Topography Coding Guidelines: Lymphoma

- Topography coding guidelines for lymphoma
  - ◆ SEER Program Coding and Staging Manual (PCSM), Appendix C, page C-699

# Topography Coding Guidelines: Lymphoma

- When a single lymph node or a single lymph node chain is involved, use the histology code describing that lymph node.

## Example:

CT shows a single enlarged cervical lymph node. This is confirmed by pathology.

Code to C77.0, lymph nodes of head, neck and face

# Topography Coding Guidelines: Lymphoma

- If the lymphoma involves multiple lymph node chains with the same topography number, code to that site.

## Example:

Facial (C77.0) and submandibular (C77.0)  
lymph nodes

Code to C77.0, lymph nodes of the head and  
neck

# Topography Coding Guidelines: Lymphoma

- When multiple lymph node chains are involved (different topography codes) at the time of diagnosis, code to C77.8.

## Example:

Positive cervical (C77.0) and mediastinal (C77.1) lymph nodes

Code to C77.8, lymph nodes of multiple regions

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# Topography Coding Guidelines for Lymphoma

- If a lymphoma is extranodal, code the organ of origin

## Example:

Small intestine resection pathology shows malignant lymphoma; no other pathologic or clinical disease.

Code to C17.9, small intestine

# Topography Coding Guidelines for Lymphoma

- If a lymphoma is present both in an extranodal site and in that organ's regional lymph nodes, code the extralymphatic organ as the primary site.

# Topography Coding Guidelines for Lymphoma

- (Continued)

**Example:**

**Stomach resection pathology:  
malignant lymphoma of gastric  
antrum; 1 of 2 gastric lymph nodes  
shows malignant lymphoma**

**Code to C16.3, gastric antrum**

# Topography Coding Guidelines for Lymphoma

- (Continued)

*Exception:* If the lymphoma in the extranodal site is a direct extension from the regional lymph nodes, assign primary site to the lymph node chain

# Topography Coding Guidelines for Lymphoma

- (Continued)

**Example:**

**Stomach resection pathology:  
malignant lymphoma of gastric antrum  
involves and extends directly from  
gastric lymph nodes**

**Code to C77.2, gastric lymph node**

# Topography Coding Guidelines for Lymphoma

- If the lymphoma is present in extralymphatic organs and non-regional lymph nodes, consult the physician to determine primary site. If primary site cannot be determined, code lymph node, NOS (C77.9).

# Topography Coding Guidelines for Lymphoma

- (Continued)

**Example:**

**Stomach resection pathology:  
malignant lymphoma of gastric antrum.**

**Biopsy of cervical lymph node:  
malignant lymphoma**

**Code to C77.9, lymph node, NOS**

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# Topography Coding Guidelines for Lymphoma

- If the primary site is unknown or not given:
  - a. Code retroperitoneal lymph nodes if described as retroperitoneal mass
  - b. Code inguinal lymph nodes if described as inguinal mass
  - c. Code mediastinal nodes if described as mediastinal mass

# Topography Coding Guidelines for Lymphoma

- (Continued)

- d. Code mesenteric lymph nodes if described as mesenteric mass
- e. If primary site is unknown code lymph nodes, NOS (C77.9)

*Exception:* Code unknown primary site (C80.9) when there is no evidence of lymphoma in lymph nodes or physician suspects extranodal lymphoma

# Topography Coding Guidelines for Lymphoma

- (Continued)

**Example:**

**CT of abdomen: retroperitoneal mass;  
suspect malignant lymphoma**

**Code to C77.2, retroperitoneal lymph  
nodes**

# Malignant Lymphoma Histology

- B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (bcll/sll)
  - ◆ If solid tumor is present code to lymphoma (9670/3)
  - ◆ If solid tumor is not present code to leukemia (9823)

# Malignant Lymphoma Histology

## ■ Hodgkin Disease

- ◆ Usually occurs in individuals between 15-35 or those over 50
- ◆ Characterized by the presence of Reed Sternberg cells
  - ◆ CD30 and CD15 positive, usually negative for CD20 and CD45
  - ◆ Derived from B lymphocytes

# Malignant Lymphoma

- **Non-Hodgkin Lymphoma**
  - ◆ A term that includes many different types of lymphoma

# Cutaneous T-Cell Lymphoma (CTCL)

- Form of Non Hodgkins Lymphoma
- Code to Skin (C44.\*)
- Treatment
  - ◆ PUVA
  - ◆ Topical or Systemic Chemotherapy
  - ◆ Radiation
  - ◆ Interferon

# Multiple Primary Rules

- SEER “Definitions of Single and Subsequent Primaries for Hematologic Malignancies”
  - ◆ Assign both histologies ICD-O-3 Codes
  - ◆ S indicates a single primary
  - ◆ D indicates a subsequent primary

# Quiz

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# Staging

# Lymphoma

- Three categories of lymphoma
  - ◆ Nodal
  - ◆ Extranodal
  - ◆ Extralymphatic

# Extranodal Lymphatic Sites

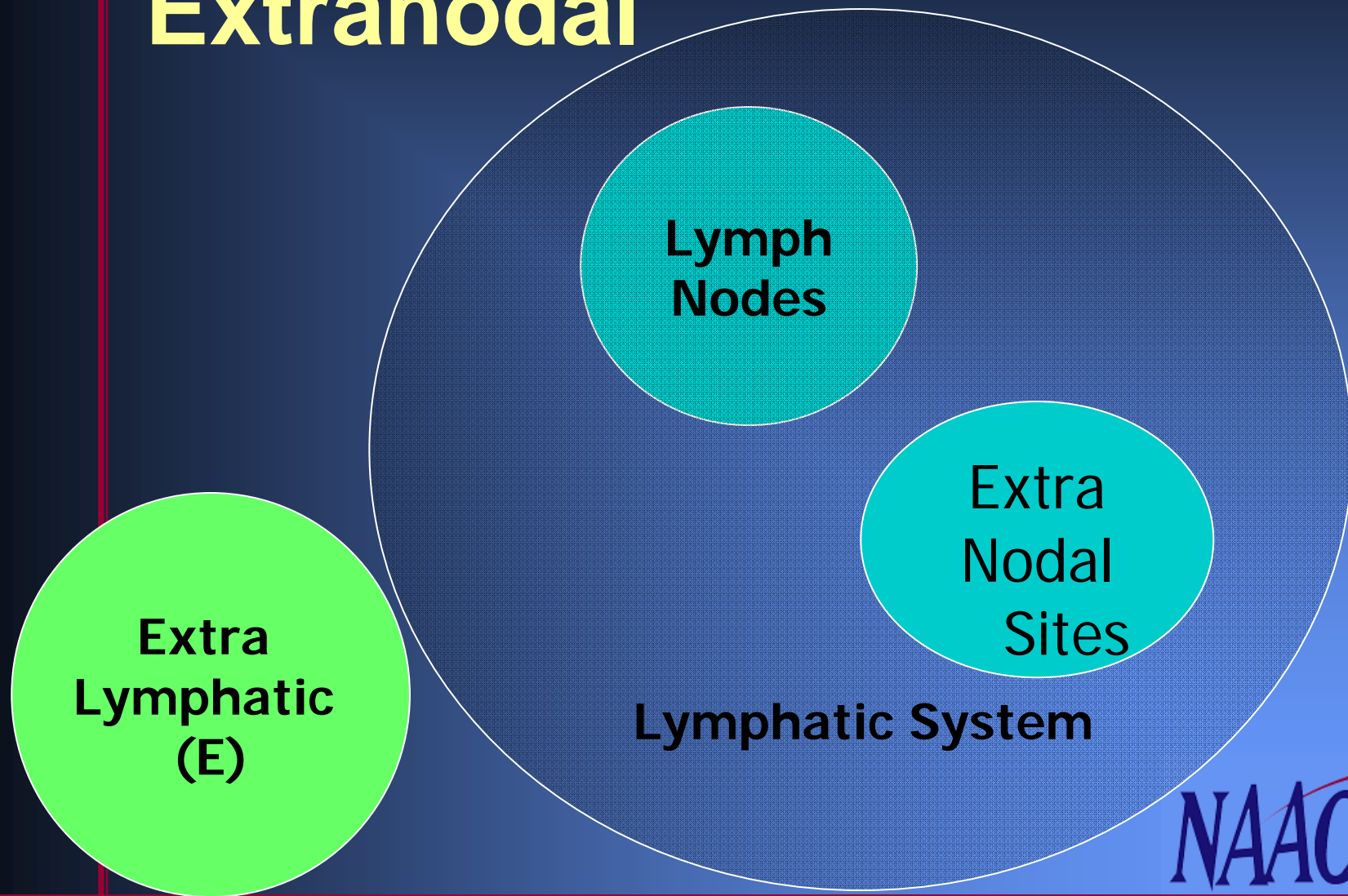
- Spleen (C42.2)
- Thymus Gland (C37.9)
- Lingual Tonsil (C02.4)
- Palatine Tonsil (C09.9)
- Waldeyers's ring (C14.2)
- Peyer's patches (C17.2)
- Lymphoid nodules of the appendix (C18.1)

# Common Extralymphatic Sites

- ◆ Stomach
- ◆ Small Intestine
- ◆ Uterus
- ◆ Bone
- ◆ Brain
- ◆ Breast
- ◆ Large Intestine
- ◆ Others

*These sites are designated by an “E” in the stage group*

# Extralymphatic vs Extranodal



# Bilateral Lymph Node Regions

- **Bilateral**
  - ◆ **Cervical**
    - ◆ cervical, supraclavicular, occipital, preauricular
  - ◆ **Infraclavicular**
  - ◆ **Axillary**
  - ◆ **Pelvic**
  - ◆ **Inguinal/femoral**
- **If both sides are involved, count as two lymph node regions**

# Axial Lymph Node Regions

- Axial
  - ◆ Mediastinal
  - ◆ Hilar
  - ◆ Para-aortic
  - ◆ Mesenteric
- Each counts as 1 lymph node region
- Extranodal lymphatic sites each count as 1 region
  - ◆ Spleen, thymus, tonsils, Peyer's patches

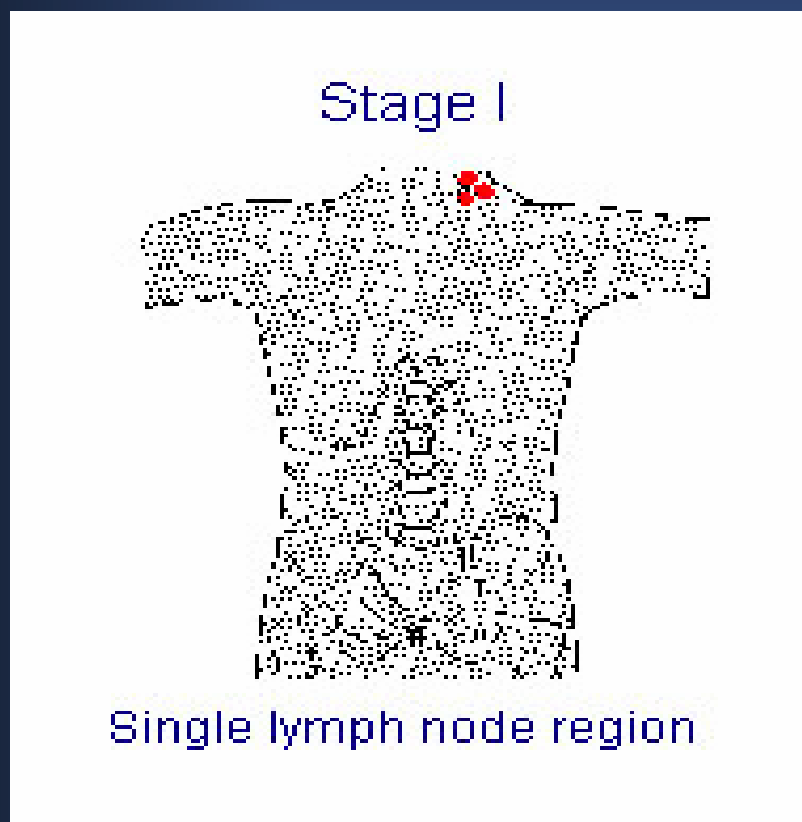
# Other Lymph Node Regions

- Internal mammary \*
- Epitrochlear \*\*
- Popliteal \*\*
- Occipital \*
- Submental \*
- Preauricular \*\*

\*Count as one lymph node region

\*\*Count as two lymph node regions if both sides are involved

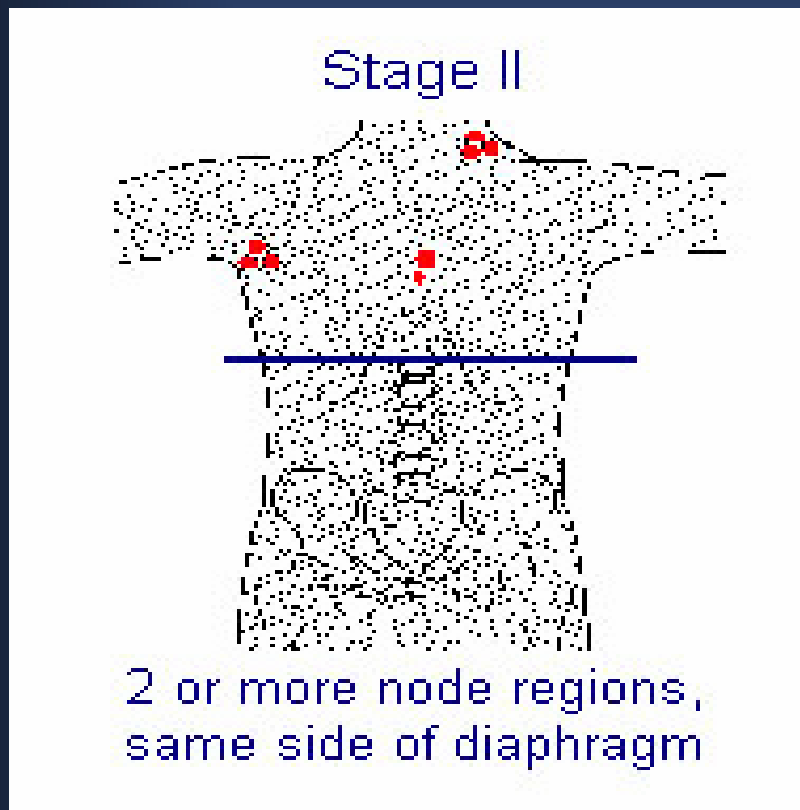
# Stage



- Stage I
  - ◆ Single Lymph Node Region
- CS Codes 10-12

Image source: SEER Training Website

# Stage

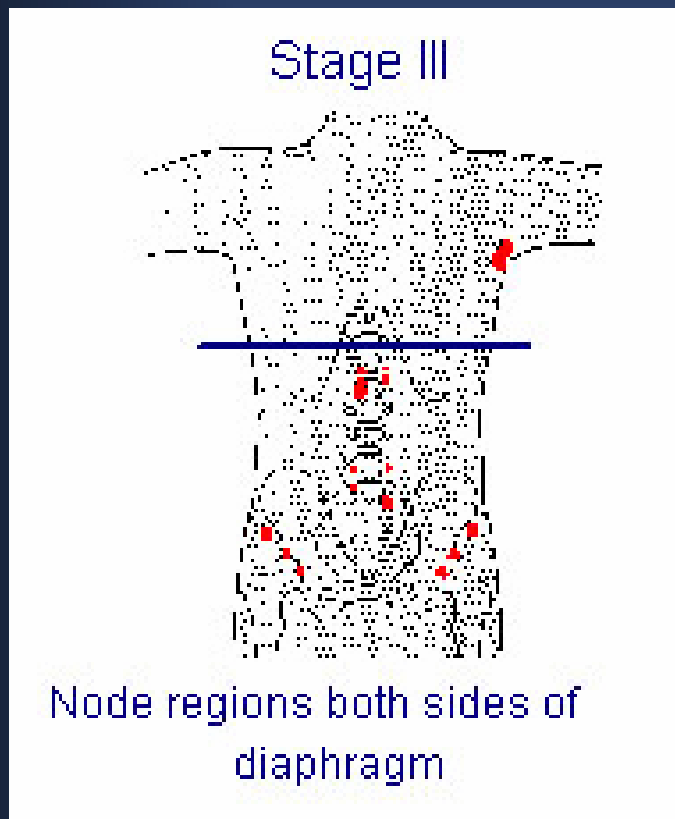


- Stage II
  - ◆ Two or more node regions on the same side of the diaphragm
- CS Codes 20-23

Image source: SEER Training Website

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# Staging



- Stage III
  - ◆ Nodal involvement on both sides of the diaphragm
- CS Codes 30-33

Image source: SEER Training Website

# Staging

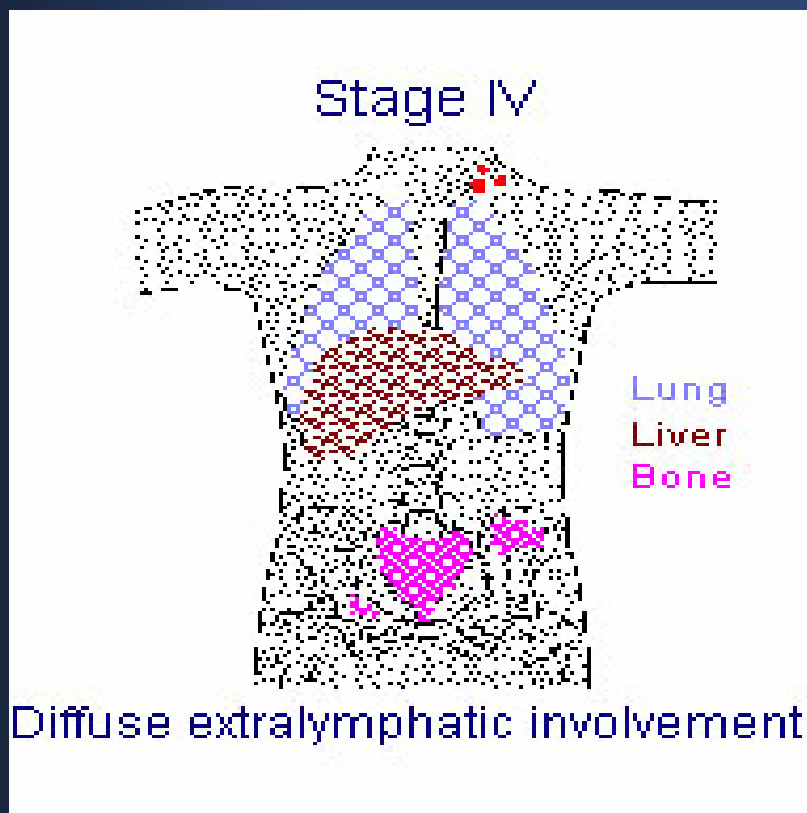


Image source: SEER Training Website

- Stage IV
  - ◆ Diffuse or disseminated involvement of one or more extralymphatic organs or any involvement of the liver, bone marrow or nodular involvement of the lungs
- CS Code 80

# Defining Lymph Node Involvement

- Clinical enlargement (without other explanation such as infection)
- Pathologic diagnosis
- Imaging: nodes larger than 1.5 cm

# “E” Suffix

- CS Extension codes 11, 21, 31
- Presenting in extralymphatic site(s)
  - ◆ Stomach, brain, etc.

# “E” Suffix Examples

- **Stage IE: primary parotid lymphoma involving entire gland that undergoes curative surgery**
- **Stage IIE: primary lung lymphoma with hilar and mediastinal disease (presenting as 2 masses)**
- **Stage IIE: mediastinal lymph nodes with direct extension to lung**

# Spleen Involvement ("S" Suffix)

- CS Extension codes 12, 22, 32
- Unequivocal palpable splenomegaly
- Equivocal palpable splenomegaly with radiologic confirmation
- Radiologic enlargement AND multiple focal defects (not cystic or vascular)

# Determining Stage IV Disease

- Site of origin
  - ◆ Stomach, colon, brain, uterus
    - ◆ Most likely extralymphatic
  - ◆ Bone, lung
    - ◆ Most likely Stage IV
  - ◆ Liver, bone marrow, cerebrospinal fluid, pleura
    - ◆ **ALWAYS** stage IV

# Lymphoma -- CS TS/Ext Eval

- Pathologic staging (eval code 3) only when staging laparotomy is done (rarely)
- Unique code structure
  - ◆ 0 No staging laparotomy
  - ◆ 3 Staging laparotomy
  - ◆ 8 Autopsy
  - ◆ 9 Unknown

# Site-Specific Factor (SSF) 1 Associated with HIV/AIDS

- 001 Yes/present
- 002 No/not present
- 999 Unknown; insufficient information; not documented

Code as 999 rather than 002 if AIDS status is not documented



# SSF 2

## Systemic Symptoms

- 000 No B symptoms (asymptomatic)
- 010 Any B symptoms
- 020 Pruritis (recurrent and unexplained)
- 030 B symptoms plus pruritis
- 999 Unknown; not documented

# SSF 3 International Prognostic Index (IPI) Score

Code	Description	Risk Group
000	0 Points	Low
001	1 Point	Low
002	2 Points	Low-Intermediate
003	3 Points	Intermediate High
004	4 Points	High
005	5 Points	High
999	Unknown, Insufficient Information, Not Documented	

# SSF 3

## IPI Score

<b>Good prognostic factors</b>	<b>Adverse prognostic factors <i>(count 1 point for each factor below)</i></b>
<b>Age below 60</b>	<b>Age 60 and above</b>
<b>Stage I and II</b>	<b>Stage III and IV</b>
<b>No more than one area of lymphoma outside of nodes</b>	<b>More than one area of extranodal involvement</b>
<b>Performance status - able to function normally</b>	<b>Performance status - needs a lot of help with daily activities</b>
<b>Serum LDH is normal</b>	<b>Serum LDH is elevated</b>

# Quiz

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# First Course Treatment

## Lymphoma

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# First Course Treatment

- Intended to affect tumor by
  - ◆ Modification
  - ◆ Control
  - ◆ Removal
  - ◆ Destruction
- Includes curative and palliative treatment

# Surgical Procedure of Primary Site: Lymphoma

- Lymph nodes
  - ◆ FORDS page 283
- Site-specific codes for extranodal sites
  - ◆ Example: Patient has partial gastrectomy; path documents primary lymphoma of stomach
    - ◆ Assign code 30 from surgical procedure of primary site codes for stomach

# Surgical Procedure of Primary Site: Lymph Nodes

## ■ Code 00

- ◆ None; no surgery of primary site; autopsy only

## ■ Code 19

- ◆ Local tumor destruction or excision, NOS
  - ◆ Principally for cases diagnosed prior to 1/1/2003

# Surgical Procedure of Primary Site: Lymph Nodes

- Codes 15

- ◆ Local tumor destruction without pathology specimen

- Code 25

- ◆ Local tumor excision, NOS

# Surgical Procedure of Primary Site: Lymph Nodes

- **Excisional biopsy of a lymph node**
  - ◆ If the intent of the procedure was diagnostic, assign code 02 under Diagnostic Staging Procedure
  - ◆ If the intent was treatment, use code 25 for Surgical Procedure of Primary Site

# Surgical Procedure vs. Diagnostic Staging Procedure

- **Example 1: Patient with hilar and mediastinal lymphadenopathy presents for mediastinoscopy; lymph node removed and found to be lymphoma**
  - ◆ **Assign code 02 for diagnostic staging procedure**

# Surgical Procedure vs. Diagnostic Staging Procedure

- **Example 2: Patient with palpable cervical lymph node presents for excisional biopsy; staging workup failed to reveal any additional disease**
  - ◆ **Assign code 25 for surgical procedure of primary site**

# Surgical Procedure of Primary Site: Lymph Nodes

- Code 30 - 32
  - ◆ Lymph node dissection
  - ◆ Code 30: NOS
  - ◆ Code 31: One chain
  - ◆ Code 32: Two or more chains

# Surgical Procedure of Primary Site: Lymph Nodes

- Code 40 - 42
  - ◆ Lymph node dissection  
PLUS splenectomy
  - ◆ Code 40: NOS
  - ◆ Code 41: One chain
  - ◆ Code 42: Two or more chains

# Surgical Procedure of Primary Site: Lymph Nodes

## ■ Code 50 - 52

- ◆ Lymph node dissection and partial/total removal of adjacent organs
- ◆ Code 50: NOS
- ◆ Code 51: One chain
- ◆ Code 52: Two or more chains

# Surgical Procedure of Primary Site: Lymph Nodes

## ■ Code 60 - 62

- ◆ Lymph node dissection and partial/total removal of adjacent organs PLUS splenectomy
- ◆ Code 60: NOS
- ◆ Code 61: One chain
- ◆ Code 62: Two or more chains

# Surgical Procedure of Primary Site: Lymph Nodes

- Code 90
  - ◆ Surgery, NOS
- Code 99
  - ◆ Unknown if surgery performed

## Scope of Regional Lymph Node Surgery Codes\*

Code	Label
0	None
1	Biopsy or aspiration of regional LNs, NOS
2	Sentinel LN biopsy
3	Number of regional LNs removed unknown
4	1-3 regional LNs removed
5	4 or more regional LNs removed
6	Sentinel biopsy and code 3, 4, or 5 at same time or timing not stated
7	Sentinel biopsy and code 3, 4, or 5 at different times
9	Unknown



# Scope of Regional Lymph Node Surgery

- Patient with inguinal lymphadenopathy; lymph node dissection removes 7 inguinal nodes with 3 positive for lymphoma; all other systems normal
  - ◆ Assign code 9 for scope of regional lymph node surgery

# Scope of Regional Lymph Node Surgery

- Patient has hemiglossectomy and modified radical neck dissection; lymphoma of lingual tonsil and 3/7 submandibular lymph nodes
  - ◆ Assign code 5 for scope of regional lymph node surgery

## Surgical Procedure/Other Site Codes\*

Code	Label
0	None
1	Nonprimary surgical procedure performed
2	Nonprimary surgical procedure to other regional sites
3	Nonprimary surgical procedure to distant lymph nodes
4	Nonprimary surgical procedure to distant site
5	Combination of codes
9	Unknown



# Surgical Procedure/Other Site

- Patient with primary stomach lymphoma has partial gastrectomy followed by excision of cervical lymph node; path shows lymphoma
  - ◆ Assign code 3, nonprimary surgical procedure to distant lymph nodes for surgical procedure/other site

# Radiation Therapy

## Radiation Treatment Volume

- 00: No radiation treatment
- 01-60: Specific site
  - ◆ 22: Mantle
  - ◆ 23: Lower extended field
    - ◆ Middle (para-aortic) field
    - ◆ Pelvic field
  - ◆ 39: Inverted Y
    - ◆ Middle and pelvic fields
  - ◆ 60: Lymph node region, NOS
- 98: Other
- 99: Unknown

# Radiation Therapy

- **Regional treatment modality (FORDS page 155)**
  - ◆ Type of regional radiation dose delivered to the primary volume (area) of interest
- **Boost treatment modality (FORDS page 159)**
  - ◆ Type of radiation provided to a smaller volume within the same volume as regional radiation

# Regional and Boost Treatment Modality

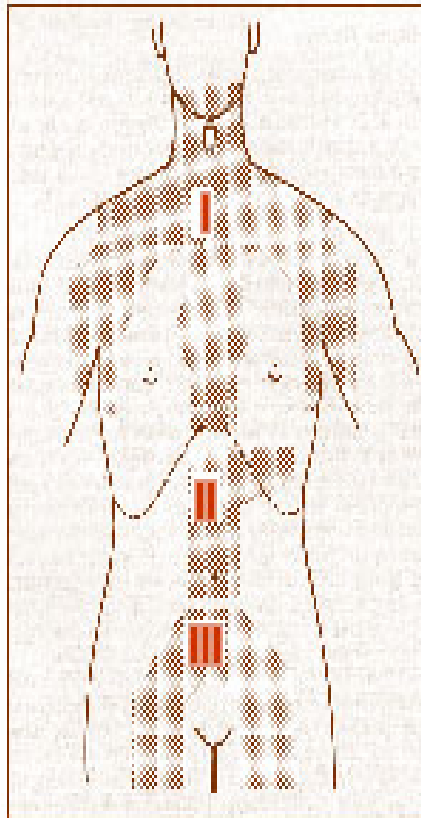
## ■ Lymphoma

### ◆ External beam radiation

- ◆ Codes 20 – 30: Orthovoltage, cobalt, photons, electrons, or neutrons
- ◆ Code 31: Intensity modulated radiation therapy (IMRT)
- ◆ Code 32: Conformal radiation

# External Beam Radiation

## Radiation Therapy for Lymphoma



- I Mantle field
- II Spade field
- I+II Subtotal nodal irradiation
- III Inverted Y
- I+III Total nodal irradiation

- Radiotherapy uses radiation, such as X-rays, to kill non-Hodgkin's lymphoma cells or to slow down their growth and development

# Radioisotopes

- **Assign code 60, radioisotopes, NOS**
  - ◆ **Drugs such as Bexxar and Zevalin combine the cell targeting ability of a monoclonal antibody with the additional cell killing ability of a radioactive particle, or radioisotope, called yttrium-90**

# Chemotherapy

- Low grade non-Hodgkin lymphoma
  - ◆ Stage 1 or 2
  - ◆ Single nucleoside
  - ◆ Single alkylating agent
  - ◆ Alkylating agent plus Prednisone

# Chemotherapy

- **Low grade non-Hodgkin lymphoma**
  - ◆ Stage 3 or 4
  - ◆ Single-agent chemotherapy
  - ◆ Combination chemotherapy with nodal radiation
    - ◆ CVP

# Chemotherapy

## ■ Fast-Growing NHLs

High grade NHL is treated with aggressive, combination chemotherapy regimens such as:

- ◆ CHOP + R
- ◆ CHOP
- ◆ BACOD
- ◆ MACOP-B
- ◆ EPOCH

See the National Cancer Institute for additional information  
<http://www.cancer.gov/cancertopics/pdq/treatment/adult-non-hodgkins/HealthProfessional/page8>



# Chemotherapy

- Rituxan

- ◆ Code as chemotherapy per SEER RX



# Hormone Therapy

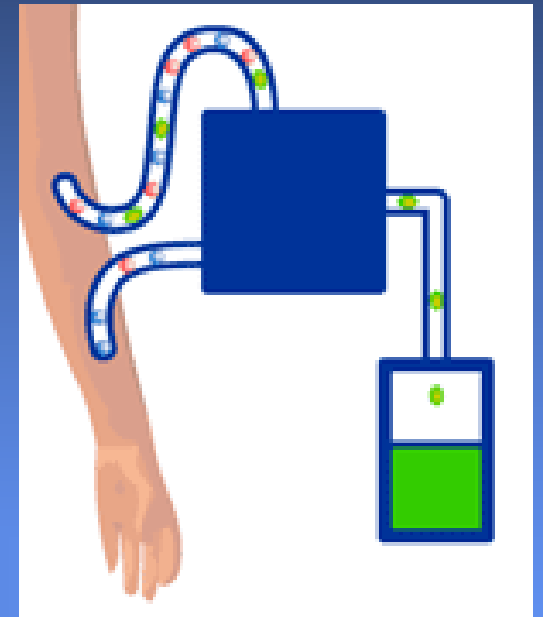
- Code hormonal agents given with chemotherapy regimens in the hormone therapy data item
  - ◆ Prednisone (ACVBP, CHOP, CNOP, EPOCH, MOCOP-B, MOPP)
  - ◆ Halotestin (EPOCH)

# Immunotherapy

- **Monoclonal antibodies (MABs or MOABs)**
- **Vaccines**
- **T-cell immunotherapy**

# Hematologic Transplant

- **Bone marrow transplant**
  - ◆ Code 11: autologous
  - ◆ Code 12: allogenic
- **Peripheral blood stem cell transplant**
  - ◆ Code 20



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# Exercises

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