This is part 1 of a 2-part presentation. This presentation will discuss certain points from the Hematopoietic Coding manual and discuss the rules for assigning the CS data items. You should follow along in your ICD-O-3, Hematopoietic Coding Rules and CS manuals.

Sources:

Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual
ICD-O-3
Collaborative Stage Manual
American Cancer Society Website
The next few slides are a review of some of the points from the Hematopoietic Coding Rules manual. It cannot be emphasized enough that the points in this presentation are not meant to stand alone. You MUST use the Hematopoietic Coding Manual. The manual will tell you exactly which code to assign.

This is only a review of certain points that are needed to carry over to assigning the CS data items correctly. Some points are included to show you how this information is listed in the ICD-O-3 manual to help you become more familiar with the contents of the ICD-O-3.
Lymphomas range from 9590 – 9729 in the numerical section of the ICD-O-3. In the alphabetic index, Lymphoma NOS and Non-Hodgkin lymphoma can be found under the ‘Lymphoma’ heading in the L section of the alphabet. Hodgkin Disease is more common in young adults. These can be found listed under ‘H’ in the alphabetic section.

The histologies listed on the slide are the current ranges and groupings for the lymphomas. These are the histologies that are included in this presentation.
What Is Lymphoma?

• Malignancy of lymphoid cells
• Responsible for immune response to infections
• Can occur in any site in which contains lymphoid tissue or to which lymphocytes (type of white blood cell) travel

An important point to remember regarding lymphoma is that it can arise in any site. It can arise anywhere lymphatic tissue is found – even the brain. Because lymphoma usually occurs in the lymph nodes, when it does arise in another site, it can often cause us to question if it is the primary site or if it is metastatic to that site from the lymph nodes.

Lymphocytes are the main type of white blood cells found in lymphoid tissue. They are either B-cell or T-cell. Lymphoma can arise in any site in which lymphocytes travel.
Malignant Lymphoma is an umbrella term for Non-Hodgkin Lymphoma and Hodgkin Disease. However, when you hear the term “lymphoma” used, without any other details, it is usually referring to Non-Hodgkin Lymphoma (NHL).

The cancer cells in Hodgkin disease are unique. They are called Reed-Sternberg cells and are an abnormal type of B lymphocyte that are much larger than normal lymphocytes. Note that Hodgkin disease is never extra-lymphatic or extra nodal.
Lymph is a clear, watery fluid that means clear spring water in Latin. The lymph fluid surrounds all of the body cells and flows through the lymph vessels. It is actually the waste product of metabolism in addition to our immune system cells. Proteins, fats, and other fluids that leak or drain out of the capillaries within various organs of the body are carried through the lymph system. The lymph fluid is carried into the upper chest and empties into the bloodstream in the neck via the thoracic duct and right lymphatic duct.

You can see where a cancer of any type and of any primary site has the potential to spread throughout the body, not only through the bloodstream, but through the lymphatic system as well.

Lymph nodes are the little bean size dots that you see throughout the body in these pictures. They are located all along the path of the lymph vessels (not just in concentrated areas which some diagrams depict) and are connected by the lymph vessels. They not only produce lymphocytes but filter the fluid and trap inflammatory substances and substances from cancerous lesions.

When we have an infection, the lymph nodes can become enlarged. So infections have to be ruled out when you enlarged lymph nodes.
When we think of lymphoma, we typically think of it as having a primary site in the Lymph Nodes (C77._). This is called nodal lymphoma.

However, lymphoma can either be classified as lymphatic or extra-lymphatic. Extra-lymphatic means the lymphoma did not originate in the lymph nodes or a lymphatic organ.

AJCC treats the terms nodal and lymphatic as synonymous terms. Therefore, lymphatic would include the lymph nodes. And, it treats extra nodal and extra lymphatic as synonymous terms. CS uses the terms lymphatic and extra lymphatic. For the purposes of this presentation, the CS terms will be used.

There are a few sites that are classified as lymphatic but are not what we typically think of as being lymphoid tissue.

The spleen, thymus gland, Waldeyer's ring (tonsils), Peyer's patches (ileum) and lymphoid nodules in the appendix are LYMPHATIC STRUCTURES. They contain mostly lymphoid tissue. Lymph nodes are also lymphatic tissues. The Lymphatic group of lymphomas includes the following:
- The Lymph nodes (C77._)
- Thymus (C37.9)
- Spleen (C42.2)
- Peyer’s patches (C17.2)
- Waldeyer's Ring (C14.2)
The spleen is the organ that makes healthy lymphocytes and stores healthy blood cells.

The thymus is responsible for making T-cells and becomes smaller and less active with age.

Peyers patches are aggregations of lymphoid tissue measuring a few centimeters in length that are usually found in the lowest portion of the small intestine (ileum). The terminology “Peyer’s patches” or “nodules in the appendix” must be used to be coded as lymphatic, code C17.2 or C18.1.

If the lymphoma arises in the “small intestine,” or “appendix,” code these as extra-lymphatic and assign the small intestine or appendix as the primary site code.
The adenoids, palantine tonsils and lingual tonsils make up the Waldeyer ring. The tonsils fight germs that are breathed in or swallowed. All of these lymphatic components work together to provide an immune system.

The Waldeyer ring has a specific site code of C14.2. To use the code for Waldeyer’s ring, this particular site must be stated or the record must show involvement of all of the listed sites are involved. If only one of the subsites is involved (i.e. pharyngeal tonsil), you cannot use Waldeyer’s ring.

There is not agreement on how a tonsil primary would be coded: as lymphatic or extra-lymphatic. For this reason, the edits have been relaxed to allow any of the tonsil codes to be coded either way. You should confer with your pathologist or physician when you have a tonsil primary for lymphoma if they would consider it a lymphatic or extra-lymphatic and code what they say.
The extra-lymphatic lymphomas are those that occur in an organ, including the skin. On this slide are a few examples of some of the more common extra-lymphatic sites in which lymphoma can arise. While they aren’t lymphoid structures, they do contain lymphocytes and therefore can be the site where the lymphoma starts. Therefore, any organ can be involved by lymphoma.

If the bone marrow is involved, before deciding that is the primary site, look elsewhere for other involvement. Lymphomas metastasize frequently to the bone marrow. It is possible to have a bone marrow primary with a lymphoma, but not very common. Specific rules are included in the Hematopoietic Coding Manual.

Hodgkin lymphoma rarely presents in a extra-lymphatic; whereas, non-Hodgkin extra-lymphatic lymphomas account for approx 25% of the cases.
For nodal lymphomas (those arising in the lymph nodes), another factor in determining the primary site is to determine how many lymph node chains are involved. The chains are divided into 6 categories in the ICD-O-3. Look at pages 64-65 in the ICD-O-3. Notice that there are many different areas of lymph nodes that fall into each of these 6 chains.

The chains are also included in Appendix C: Lymph Node/Lymph Node Chain Reference Table in the Hematopoietic Coding Manual. When reviewing the rules in the Hematopoietic Coding Manual, the rules say to refer to appendix C. You should use Appendix C and not the ICD-O-3. However, it is important to understand the information provided to you in the ICD-O-3. And, it will help you understand how the rules and appendix were developed.

Reminder: You MUST use the Hematopoietic Coding Manual. The manual will tell you exactly which code to assign.
The site code that is to be assigned is determined by several factors. This slide contains a summary of some of the primary factors including whether the site is nodal or extranodal and the number of lymph node chains involved. In the next few slides, we will discuss each of these more detail.

These factors are based on Module 7: Primary Site Rules for Lymphomas Only (PH25-PH37) and Appendix C in the Hematopoietic Coding Manual. You may want to follow along in your ICD-O-3 and the Hematopoietic Coding Manual to see how these are listed in each.
If it is clear that a specific lymph node chain is the primary site, code to that lymph node chain. Hopefully, through the diagnostic workup, the physicians will be able to make the determination that only one LN chain was involved.

For example, the only area of involvement is the Parotid lymph nodes. The site will be C77.0. There may be more than one lymph node in the parotid chain involved, but all of them must be in the same chain for that ICD-O-3 site code.

Another example is facial and submandibular lymph nodes. Both have the same site code of C77.0. However, this is two different lymph node chains according to staging. The site code that should be assigned is still C77.0, lymph nodes of head and neck, because only one lymph node chain according to ICD-O-3 is involved.
When a mass is identified as retroperitoneal, inguinal, mediastinal, or mesentery and is consistent with lymphoma, code to the specific lymph node site: retroperitoneal (C77.2), inguinal (C77.4), mediastinal (C77.1), or mesentery (C77.2).

The presumption is that no other lymph node regions are involved. This does not apply to systemic disease.
The ICD-O-3 coding rule for overlapping lesions is a little different for lymphomas. For lymphomas, C77.8 indicates that there are multiple lymph node regions involved.

For C77.8 to apply, the lymph nodes involved must have different site codes when you look them up in Appendix C.

For example, if the axillary (C77.3) lymph nodes and inguinal (C77.4) lymph nodes are involved, the site would be C77.8 because axillary and inguinal have different topography codes.
If it is not clear what the primary site is, and it is known to have risen in the lymph nodes, code to C77.9, lymph nodes, NOS.

The key here is that the lymph nodes are known to be involved. If the record only states that the patient has lymphoma, nos without specific details, code to C77.9.
If there are multiple areas of involvement, they may choose to biopsy a particular LN because it is the most accessible. Do not automatically code to the lymph node that was biopsied (unless it is the only chain involved and it is stated that this it is the primary site, of course. That was the first rule we discussed).

Also, you should code to C77.9 if a LN biopsy is done, and the pathology was consistent with lymphoma, but it is not clear what the primary site is. Because it was a lymph node that was biopsied, we know that the lymph nodes are involved. The site code of C80.9 (unknown primary) would not be appropriate in this situation. Because we don’t know exactly what the primary site is (one chain, multiple chains, lymph nodes + an extranodal sites, etc.) we should code this to C77.9 (lymph nodes, nos).
If there is involvement of an extra-nodal site (the site is not the lymph nodes), and there is no nodal involvement of any kind, code to the extra-nodal site. For example, C16 for Stomach or C14.2 for Waldeyer’s ring.

If there is nodal involvement in the area, it should be stated in the medical record that the origin was an extra-nodal site.
When lymphoma presents in an extranodal site and in the regional lymph nodes for that extranodal site, code the Primary Site to the extranodal site. The typical disease process is that lymphoma can spread from an extranodal organ to its regional lymph nodes. It cannot metastasize from the regional lymph node back to the extranodal organ. The exception to this would be if the lymph nodes presented as one large mass that extended into the regional organ.

Physiologically, lymphoma cells in regional lymph nodes do not "back-flow" into the extralymphatic organ to involve it secondarily. As a result, the primary site is usually considered to be the extralymphatic organ with regional lymph node involvement. Do not be afraid to code an extralymphatic site as primary when that site and its regional nodes are involved.

Example: A patient with a submandibular lymphoma and involved nodes undergoes a salivary gland excision and a modified radical neck dissection yielding 100 nodes. Code the Primary Site to C08.0 [submandibular gland] and use the surgery code schemes that apply to that site (Parotid and Other Unspecified Glands).

If the lymph nodes are not considered regional to the extra-nodal involved site and the primary site cannot be determined, code the primary site to C77.9 [Lymph node, NOS]. This is consistent with the information on the previous slide.

Of special note: Brain, spinal cord, bone marrow, cartilage do not have regional lymph nodes.
If it is suspected that the lymphoma is extra-nodal and no site code is given, code to an Unknown Primary Site (C80.9).

There can be nodal involvement. The key is where it is thought the lymphoma arose and if it was NOT in the LNs, then code to C80.9.

For example, if a diffuse large B-cell lymphoma is found in the femur and in the soft tissue of the anterior chest wall but all CT scans are negative for lymphadenopathy, code the Primary Site field to C809 [Unknown primary site]. Diffuse large B cell lymphoma can be either nodal or extranodal. The case described above is likely extranodal because there is no evidence of lymph node involvement. Because there are multiple areas of involvement and the extranodal site of origin is unknown, code the Primary Site to C809.

This rule, that allows us to use the C80.9 site code, took effect with the implementation of the ICD-O-3 and with cases diagnosed after 1/1/2001.
Always document the primary site (any site), but most especially when there is involvement of both an extra-nodal site and lymph nodes.

You will have the documentation readily available to defend your abstracting and it will clearly document that this is an extranodal lymphoma with regional lymph node involvement and not a stage 4 nodal lymphoma.

In addition, always document why you are using Unknown Primary as your site code, not just for lymphoma cases but all of your “unknowns”.
Because we don’t see these too often, here’s a coding tip. These two conditions are coded to the skin (C44).
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>T-Cell (pre-T cell, T-precursor)</td>
</tr>
<tr>
<td>6</td>
<td>B-Cell</td>
</tr>
<tr>
<td>7</td>
<td>Null Cell (primarily for leukemias)</td>
</tr>
<tr>
<td>8</td>
<td>NK Cell (uncommon in U.S.)</td>
</tr>
<tr>
<td>9</td>
<td>Cell type not determined, not stated or not applicable.</td>
</tr>
</tbody>
</table>

For hematopoietic primaries, the 6th digit of the histology code is the immunophenotype. The immunophenotype denotes the cell's lineage or “family of cells”. (For other sites, the 6th digit is the grade or differentiation of the tumor – well differentiated, moderately differentiated, etc.) Listed here are the phenotypes for lymphoma and leukemia.

B-cells transform into plasma cells and secrete antibodies that attach to the infection and causes it to die. B-cells go straight from the bone marrow into the lymph system.

T-cells are also called killer cells. They attack foreign substances by engulfing it with a toxic effect. T-cells are formed in the thymus gland.

Null cell means, non t-non b cell and is used primarily for leukemias.

NK cell refers to “Natural killer cell" and isn’t seen too much in the US…..mainly seen in the Mediterranean area and Africa.

There is not a combination code for when both T-cell and B-cell are included in the diagnosis. And, there has not been any decision that we can give one priority over the other. Coding such a case as a pure B cell or a pure T cell would misrepresent the phenotype. So, if there is a combination of B & T cell, the only option we have to code to 9. There is a rule for this in the Hematopoietic Coding Manual.
The rules in the Hematopoietic Coding Manual stated that only codes 5-9 can be used for hematopoietic primaries. It also stated not code references to grade. In some instances, the term “grade” does not imply differentiation and should not be used to code the 6th digit of the morphology code. We talked about this with the intraepithelial neoplasias in the ICD-O-3 presentation. The same applies for lymphomas. Pathologists may use the term as a synonym for “type” or “category”.

The grades of follicular lymphoma and nodular sclerosing Hodgkin lymphoma are actually types or categories of these diseases. On page 168, under Follicular, you’ll see grade 1, 2 or 3. Do not code these in the differentiation field (Rule G3). The rules may state that another immunophenotype code would apply. Example: left submandibular neck mass consistent with malignant lymphoma, follicular type, grade III. Assign the 6th digit as 6, not 3 or 9. Rule G6 states to assign this histology to B-cell, even if B-cell is not specifically mentioned.

For lymphomas, do not code the descriptions "high grade," "low grade," and "intermediate grade" in the Grade, Differentiation field (Rule G3). These terms refer to categories in the Working Formulation and not to histologic grade. Code the grade to 9 [cell type not determined, not stated or not applicable]. Example: retroperitoneal lymph node bx: low-grade follicular lymphoma. Again, assign the 6th digit as 6 (Rule G6).
Determining Stage
Reading Assignments

- As each data item is being discussed, you should stop and read the information in CSV2 Part II and CSV2 Part I, Section 2 for that data item including the associated notes, codes and definitions.
- Also, review the AJCC chapter for Lymphoma (Chapter 57A)

It is important that you follow along and make notes in your manual. In addition to reading the slides and the instructor’s notes, it is important that you stop and read the related sections in your manual as not every point will be discussed in detail.
If you will remember from Module 5, the hematopoietic cancers are considered histology specific – the staging scheme is guided by the histology code. Therefore, for lymphoma, whether the primary site is the lymph nodes or another organ, you should use the lymphoma staging scheme.

This means you should use the lymphoma staging scheme for all lymphomas, regardless of primary site - even for lymphomas of the brain or of an unknown primary site. Also, Non-Hodgkin and Hodgkin lymphoma use the same CS coding schema.

The one exception is for lymphomas of the ocular adnexa. A lymphoma of this primary site has its own staging scheme. In the PDF files for the schema groups, this is grouped with the Ophthalmalic Sites PDF.
Differentiating lymph node regions for staging and lymph node chains for ICD-O-3 coding can be confusing. Remember to keep the rules separate. When assigning the stage, use the staging rules. When assigning the ICD-O-3 code, use the ICD-O-3 rules.

For staging, the AJCC Manual defines the LN regions. These are also defined in the 2010 Hematopoietic and Lymphoid neoplasms rules. There is an excellent table in the Hematopoietic and Lymphoid Neoplasm Case Reportability and Coding Manual (Table C1: Lymph Node/Lymph Node Chain Reference Table). You may want to print this out and place with your CSv2 lymphoma schema.

Once you know the LN regions involved, then you look it up in the ICD-O-3 to determine the primary site code. Lymph node chains are subcategories of LN regions. The number of chains involved affects the site code. The number regions involved affects the stage. It is important to keep all of these factors in mind.

For example, in the previous slide, intrathoracic LN (C77.1) were listed as above the diaphragm.

The ICD-O-3 code for intrathoracic LNs includes both mediastinal and hilar lymph nodes. Mediastinal and hilar lymph nodes have the same primary site of C77.1. But, according to AJCC, these are considered two separate LN regions for the purpose of staging. You will see where this is important when we talk about the CS Extension data item.
There are many lymph node regions. Knowing whether there is involvement of lymph nodes on one or both sides of the diaphragm is important.

You can find a complete list of LN regions in the AJCC manual. In this drawing, the different colors represent a different lymph node region. Notice that for C77.1, Mediastinal and Hilar are 2 separate lymph node regions for the purposes of staging even though they have the same primary site code.

Appendix C in the Hematopoietic Coding Manual also compares the ICD-O-3 lymph node chain to the AJCC lymph node region.
Lymph Node Regions

Each color represents ONE lymph node region for staging purposes.

Slide 30

This drawing shows the lymph node regions below the diaphragm. Again, notice that mesenteric and para-aortic are two different lymph node regions even though they have the same primary site code.
Laterality is also an important factor for staging of lymphomas.

Axial nodes are those that are centrally located. Each “axial” or central region counts as ONE LN region.
Bilateral LNs are those that are found on both sides of the body. For example, you can have either R axillary involvement, L axillary involvement, or both.

If LNs on both sides of the body are involved – even if it is the same chain with the same ICD-O-3 code, then this is counted as two LN regions being involved. The correct CS Extension code is 200.

Because they are both involving the same chain (axillary LNs) with the same ICD-O-3 code, it is considered involvement of one LN chain and therefore coded to C77.3.
Bilateral involvement of a paired **extranodal organ** is considered involvement of a single organ - as long as there is no other involvement.

You may want to make a note of that in your CS manual because you will probably not see that too often and it would be easy to forget.
Considered as Lymph Node Involvement

- Any mention of enlarged lymph nodes is indicative of involvement
  - Palpable, rubbery, enlarged, shotty, matted, visible swelling
- Clinical enlargement without explanation for cause such as infection
- Pathologic diagnosis
- Nodes larger than 1.5cm on imaging
- Caution: “rock hard” may indicate a carcinoma instead of a lymphoma

Lymphomas ONLY!!

For lymphoma, there are a few special circumstances in which the lymph nodes and spleen are considered to be involved.

These are a little different from the other solid tumors. So remember, this is for lymphomas only – not carcinomas.

Any mention of enlarged lymph nodes is considered involvement. This is often missed when reviewing the medical record. The statement "For lymphomas, any mention of lymph nodes is indicative of involvement" refers to the terms list here: Palpable, enlarged, visible swelling, shotty, lymphadenopathy. While these terms are ignored for other malignancies, they should not be ignored for lymphomas.

Of course, if they say the enlargement is due to another cause, such as an infection, then obviously you do not consider that as involvement.

AJCC also talks about liver, lung, CNS, and bone marrow involvement. You may want to copy this page and place in your CS manual in the lymphoma section.

Be careful if they describe the mass as rock hard, this may indicate a carcinoma instead of a lymphoma.
For a clinical diagnosis of splenomegaly, it depends if the splenomegaly is considered equivocal or unequivocal. Equivocal means “uncertain nature”. If there is equivocal (uncertain) palpable splenomegaly, then radiologic confirmation is also necessary before you can consider there to be lymphoma involvement of the spleen.

Note: An enlarged spleen on CT is not an indication of involvement. Nodules on the spleen must be noted.
Involvement of soft tissue adjacent to a lymphoma site does not alter the staging.

*Example:* Lymphoma of the 3rd lumbar vertebra with extension into the soft tissues surrounding the bone but without LN involvement.

CS Ext = Code 110
AJCC = Stage IE
SS2000 = Local
There are two CS data fields and 5 SSF’s that must be assigned in the Collaborative Staging System for lymphoma.

The remaining data items are either all 8’s or 9’s. Notice that the CS tumor size should be coded to 988, not 999.
CS Extension is the field that will derive the AJCC Stage group and uses the lymph node regions involved as the criteria. We have talked about what is considered a region in the previous slides. Read each code definition carefully to select the code that best describes the extent.

Review Note 1. Remember that a lymphatic structure is also considered as a lymph node region in assigning the CS Extension data item.
To assign the CS Extension, it may be helpful to ask the following questions about the case. You need to know how many LN regions are involved, if there are extralymphatic organs involved, if the spleen involved, or if there is involvement on one or both sides of the diaphragm.

Once you know that information, determining the CS Extension code is a little easier.
The diaphragm's role in staging:
- The muscle that separates the chest (thoracic) cavity from the abdomen.
- Is the dividing point for staging in lymphomas (below and above, on both sides)

The diaphragm is used in several ways:
- Same side of the diaphragm
- Opposite side of the diaphragm
- Both sides of the diaphragm

The diaphragm is usually used in distinguishing between different stages. For example, with lymphatic primaries, lymph node regions involved on the same side of the diaphragm are Stage II. In Stage III, lymph node regions are involved on both sides of the diaphragm.
Just as we did with some of the other sites, we are going to discuss the CS Extension codes based on how they are grouped. In this schema, it is based on the type of involvement. The first group of codes we will look at are those codes that are used when the primary site is a lymphatic site/structure. Remember that this includes lymph node involvement and/or involvement of lymphatic structures.

In the 2010 Hematopoietic and Lymphoid neoplasms rules, there is an appendix that lists the regions of lymph nodes. You can have involvement of two different lymph node chains that are in the same lymph node region. So, it is important to use this appendix to determine if you have one or more regions. You can also use the pictures of the regions in AJCC 7th edition.

Code 100 is used when you have involvement of only one lymph node region.

Code 200 is used when you have involvement of more than one lymph node region on the same side of the diaphragm. Code C77.8 can be used with this code (not with code 100).

Code 300 is used when you have involvement of more than one lymph node region on both sides of the diaphragm. Code C77.8 can be used with this code.

If you have involvement of a lymphatic sites AND lymph nodes, use codes 200 or 300 depending on where the lymph node regions are (above or on both sides of the diaphragm.)
The next group of codes are those that are used when the primary site is an extralymphatic organ. Extralymphatic codes are for all other sites, which are usually organs (i.e. stomach, CNS, lung, breast).

Code 110 is used when the only site involved is the primary site (the extralymphatic site). There is no lymph node involvement.

Code 210 is used when there is involvement of an extralymphatic site PLUS lymph nodes on the SAME side of the diaphragm; or there is direct extension to adjacent tissues or organs.

Code 310 is used when there is involvement of an extralymphatic site PLUS lymph nodes are on OPPOSITE sides of the diaphragm.
Even though Spleen is considered a lymphatic site, it does not use the same set of codes that the lymphatic sites use. Spleen involvement has their own set of codes.

Code 120 (Stage IS) should only be used when the ONLY organ involved is the Spleen and NO lymph nodes are involved. Do not use this code for any other site.

Code 220 (Stage IIS) states that the Spleen is involved plus lymph nodes below the diaphragm. For this code, the spleen or any other lymphatic structure can be the primary site. For example, the thymus may be the primary site and in addition to the thymus, the spleen and lymph nodes below the diaphragm are also involved.

Code 23 (Stage IIES) states that the Spleen and an extralymphatic organ are involved. In this case, your primary site should be an extra-lymphatic organ that is BELOW the diaphragm. There may or may not be involvement of the lymph nodes BELOW the diaphragm.
Code 320 (Stage IIIS) states involvement of the Spleen and nodes ABOVE or on BOTH sides of the diaphragm are involved. Your primary site should be a lymphatic site/structure, including spleen.

For code 330 (Stage IIIES), there are two different scenarios that apply:
- The Spleen AND an extralymphatic site ABOVE the diaphragm are involved. There may or may not be lymph node involvement on either side of the diaphragm.
- The Spleen AND an extralymphatic site on EITHER side of the diaphragm are involved. There is involvement of lymph nodes ABOVE the diaphragm.
- For either scenario, the primary site should be an extralymphatic site.

As you can see, it is important to pay close attention to the use of the words ABOVE, BELOW, BOTH, and EITHER side of the diaphragm. It is also important to pay attention to the use of the words LYMPHATIC and EXTRALYMPHATIC.
Code 800 is used for metastatic involvement, usually the bone marrow, liver, and nodular involvement of lung(s).

Code 800 includes several different scenarios:

- Diffuse or disseminated (multifocal) involvement of ONE OR MORE extralymphatic organ(s)/site(s) WITH OR WITHOUT associated lymph node involvement
- Multifocal involvement of MORE THAN ONE extralymphatic organ/site
- Involvement of isolated extralymphatic organ in absence of involvement of adjacent lymph nodes, but in conjunction with disease in distant sites

Remember, when you have a positive bone marrow biopsy for lymphoma, you need to check for evidence of a primary site. A primary site of bone marrow for lymphoma is possible, but it is rare. In the case that you have a bone marrow primary for lymphoma, you would use code 800 since bone marrow involvement, whether the primary site or metastatic, is systemic.

Use code 999 when you cannot determine the extension.
This table is a summary for assigning appropriate Extension codes for lymphoma.

When determining extent of disease for lymphomas your primary site determines which set of codes should be used. For lymphatic structure primary sites, the first column of codes are the ones that are applicable. For all other sites, use the second column. The last column applies when the spleen in involved, regardless of primary site.
This slide provides some examples of how various types of involvement affect the CS Extension data item.

1) Cervical and supraclavicular lymph nodes on the same side of the body. This is involvement of one lymph node region on SAME side of diaphragm.
   CS Ext: 100
   AJCC Stage: I, SS2000: Local

2) Cervical lymph nodes on both sides of the body (bilateral = two regions) This is involvement of two or more lymph node regions on the SAME side of the diaphragm.
   CS Ext: 200
   AJCC Stage: II, SS2000: Regional, NOS

3) Cervical and para-aortic lymph nodes This is involvement of lymph node regions on both sides of the diaphragm.
   CS Ext: 300
   AJCC Stage: III, SS2000: Distant

4) Cervical, supraclavicular, mediastinal lymph nodes plus the small intestine This is involvement of an extralymphatic organ PLUS involvement of lymph nodes on the OPPOSITE side of the diaphragm.
   CS Ext: 310
   AJCC Stage: III E, SS2000: Distant
These are the equivalent codes for the three staging systems for lymphoma.

Notice that AJCC TNM stage 3 and 4 are equal to Summary Stage 7 or Distant disease. There is no summary stage code of 2, 3 or 4. 1, 5, and 7 are the only valid codes for lymphoma summary stage.
The data item, CS TS/Ext-Eval, has been modified for the lymphoma schema. For lymphomas, this field relates to whether or not the patient had a staging laparotomy or was diagnosed at autopsy. Staging laparotomy is required for pathologic staging but is no longer considered a useful procedure for staging and has essentially been abandoned with new technology.
A common coding error is using code 9 for unknown when it should be coded to 0 (clinical staging basis). If CT, MRI and radiological exams are used for staging purposes, this is to be coded as 0. Even surgery or a bone marrow biopsy would still be assigned an eval code of 0. An eval code of 0 is applicable for a majority of lymphomas since they have some form of workup to establish stage (clinical). If there is no workup, code 9 for unknown would be applicable.

For example, a lymphoma of the tonsil is diagnosed. A tonsillectomy is performed, removing the entire primary site. This would not be an eval code of 3 based on this surgery alone even though the primary site is removed. The eval code would be 0. For AJCC Staging, a staging laparotomy (or autopsy) is required for a pathologic staging basis. If a staging laparotomy were performed following surgical removal, then an eval code of 3 could be assigned.

Remember that the TS/Ext eval for Lymphoma is different from all other sites. Coding a TS/Ext of 3 in Lymphoma will be the exception rather than the norm for surgical cases.
For pathological staging, a staging laparotomy (or laparoscopy) is required. This requires removal of the spleen, a liver biopsy, a lymph node biopsy and a bone marrow biopsy. Staging laparotomies are not done very often and are somewhat outdated.

According to AJCC, the use of the term pathologic staging is reserved for patients who undergo staging laparotomy with an explicit intent to assess the presence of abdominal disease or to define histologic microscopic disease extent in the abdomen. Staging laparotomy and pathological staging have been essentially abandoned as useful procedures. CS used a modified evaluation scheme for lymphomas and it applies to the CS TS/EXT-EVAL field only. The other EVAL fields are coded as “not applicable” for this schema.

There are two codes for autopsies. These codes are in alignment with all other site specific schemas. The code that applies depends on whether it was known or not that the patient had lymphoma before they died.

Remember, for lymphoma, there are only 4 eval codes for CS TS/Ext, whereas for most sites, there are 8 codes.
The data items listed on this slide are “not applicable” for Lymphoma. For the Grade Path Value/System, the only codes available are 2, 3, 4 and blank, which is why this field would be left blank instead of having a numerical code.
Site Specific Factors

- SSF 1: Associated with HIV/AIDS
- SSF 2: Systemic symptoms at diagnosis
  - A = Asymptomatic
  - B = B symptoms
    - Drenching night sweats, soaking of bedclothes
    - Unexplained fever (above 101.5 degrees F)
    - Unexplained weight loss – more than 10% of body weight in the 6 months prior to diagnosis

There are 5 SSF’s collected for lymphoma. The first two are fairly straightforward. It is important to pay attention to when you can code none vs unknown for each SSF.

For SSF 1 (HIV/AIDS), do not assume it is negative if there is no mention, you should code to unknown.

For SSF 2, if the H&P and/or clinical history, makes no mention of the “B” symptoms, you may code “none”. To code for positive B symptoms, the case must meet the listed criteria, which includes night sweats that require bed changing, fever that is not due to some other illness and weight loss of greater than 10% which cannot be explained. Pruritis, itching, can be caused by several factors. A mild cause of pruritis can be dry skin, which results in itching and is usually remedied by a localized treatment. To be considered a B symptom for Lymphoma, pruritis must be burning and intense itching that is not reactive to local treatment. This type of pruritis usually responds to chemotherapy or radiation. If just pruritis is mentioned, with no other documentation, use code 20. This will map to no B symptoms.
There are three indices/scores associated with Lymphoma:
• The International Prognostic Index (IPI, SSF3)
• The Follicular Lymphoma International Prognostic Index (FLIPI, SSF4) and
• The International Prognostic Score (for Hodgkin lymphomas, SSF5)

The International Prognostic Index was initially developed for B cell lymphomas. With CSv2, other indexes/scores have been added for more specific histologies. SSF3 is now considered to be for B cell lymphomas, although no histologies restrictions will be enforced on this SSF or the next two. The two new SSF’s are defined much more clearly as to which histologies are applicable. For all other histologies, use this SSF.

Several questions were submitted regarding how to code “low”, “intermediate” or “high” risk instead of points. These choices have been added in additional codes.

The criteria for this index is listed in the CS manual, but do not take this as a way to calculate the points yourself. Only enter the score if the physician documents it in the record. You are not required to follow up with the physician if this score is not documented. The IPI score is to be recorded as stated in the medical record. This is usually stated by the oncologist in the H&P but may also be on the AJCC TNM staging form.

The FLIPI and IPS are very similar to the IPI. The FLIPI will be for the Follicular Lymphomas. The IPS will be for Hodgkin lymphomas. Both of these SSF’s follow the same code pattern and instructions as SSF3. But, if a physician uses any of these, they may be recorded regardless of the histology and code the other two SSF’s as 999. Again, no histologies restrictions will be enforced on these SSF’s.
The International Prognostic Index was designed to further clarify lymphoma staging. The IPI predicts the risk of disease recurrence and overall survival by taking into account factors such as age, stage of disease, general health (also known as performance status), number of extranodal sites, and the presence or absence of an elevated serum enzyme named lactate dehydrogenase or LDH.

There are five factors that are important. For each factor, the 1st line indicates a good prognostic factor. The second line indicates adverse prognostic factors. For each adverse prognostic factor that the patient has, 1 is added to the total IPI. This is not something you should compute yourself. This is to be coded as recorded in the medical record by the oncologist. If it is not stated in the medical record, then assign 999 for unknown.
Here is an example of a patient with an IPI score of 5. Each one of these gets a count of 1, for a total of 5.

- Age over 60 years
- Late-stage disease (3 or 4)
- More than one extranodal site
- Poor general health
- High LDH
Based on that information, the physician will have an indication of risk and prognosis. For the patient in this example, the risk will be high for recurrence, with a 44% complete response rate; 40% chance of a relapse-free 5-year survival rate and an overall 5-year survival rate of 26%.

Of course these numbers are based on a group of individuals with the same qualities. Some individuals may do better or worse than the noted percentages. The IPI chart is only to help physicians gain additional insight for treatment and prognosis. But, as a general assumption, the higher the IPI, the worse the prognosis.
This slide contains some additional information for staging mycosis fungoides.

Mycosis fungoides is the most common form of cutaneous T cell lymphoma (CTCL). Sezary Syndrome is the aggressive leukemic form of CTCL. MF and SF have different staging systems because they do not have the same prognosis as the other skin lymphomas.

In AJCC 7th edition, MF has its own chapter (Primary Cutaneous Lymphoma). Both CS and Summary Stage 2000 also give MF its own schema. In the CSv2 manual, mycosis fungoides is listed under “musculoskeletal and skin”.

Also note that Multiple Myeloma and Plasma Cell Disorders have their own AJCC chapter (chapter 57C). And, Pediatric Lymphoid malignancies have their own AJCC chapter (chapter 57D).
Concluding remarks

- Know the difference between a lymphatic and an extralymphatic structure
- Know the difference between an ICD-O-3 lymph node chain and an AJCC lymph node region
- Pay attention to involvement above or below the diaphragm
- Use codes that are applicable based on the primary site (lymphatic or extralymphatic)

This concludes this presentation.

Please continue with Part 2.