Collecting Cancer Data: Hematopoietic

NAACCR 2011-2012 Webinar Series

August 2, 2012

1

NAACCR

Q&A

- Please submit all questions concerning webinar content through the Q&A panel.

Reminder:

- If you have participants watching this webinar at your site, please collect their names and emails.
  - We will be distributing a Q&A document in about one week. This document will fully answer questions asked during the webinar and will contain any corrections that we may discover after the webinar.

2

NAACCR

Fabulous Prizes

3

NAACCR

Agenda

- Overview
- Hematopoietic Database and Manual
- Collaborative Stage Collection System V02.04
- Treatment

4

NAACCR

Ask a SEER Registrar

- All questions about hematopoietic and lymphoid neoplasms concerning:
  - Multiple primaries
  - Primary site
  - Reportability
  - Grade
  - Any other issues related to the Hematopoietic and Lymphoid neoplasms Database and Manual
- Should go to Ask a SEER Registrar
  - http://seer.cancer.gov/seerinquiry/index.php?page=search

5

NAACCR

Ask a SEER Registrar

Members of the cancer registrar community may use this page to submit questions to SEER about coding cancer cases or about the materials for registrars distributed through the SEER site.

Note: Follow local procedures for submitting questions to your central registry when required. Your central registry will submit the question to SEER if needed.

Please note that questions about Collaborative Stage should be directed to the Commission on Cancer's CAnswer Forum.

SEER Inquiry System

Answers to coding and abstracting questions can be found in the SEER Inquiry System. Please search the system before contacting us with a question.

Submit a Question to a SEER Registrar

Questions submitted through this form will be sent to the appropriate SEER personnel. The question and answer may be added to the SEER Inquiry System for others to reference.

Choose a subject:
- ⦿ Hematopoietic Rules (database and manual)
- ○ Multiple Primary & Histology Rules
- ○ SEER*Rx - Interactive Drug Database
- ○ SEER Manual
- ○ Other

Your e-mail address: _____ *(required)*
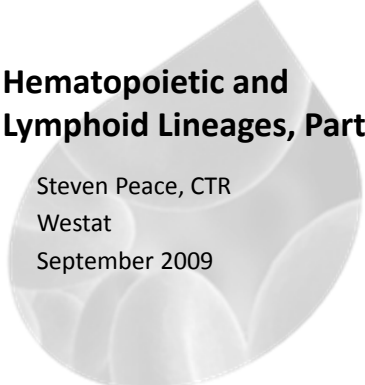☐ Send a copy of this message to yourself

Your question:

Optional contact information
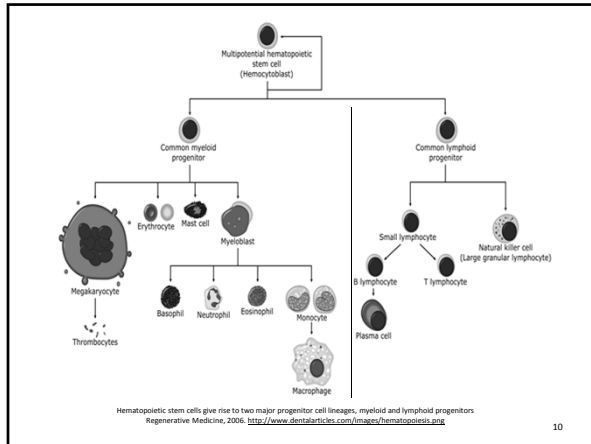Name: _____
Telephone: _____

7

[Submit Question]

OVERVIEW

8

NAACCR

Hematopoietic and Lymphoid Lineages, Part I

Steven Peace, CTR

Westat

September 2009

http://seer.cancer.gov/tools/heme/training/   9

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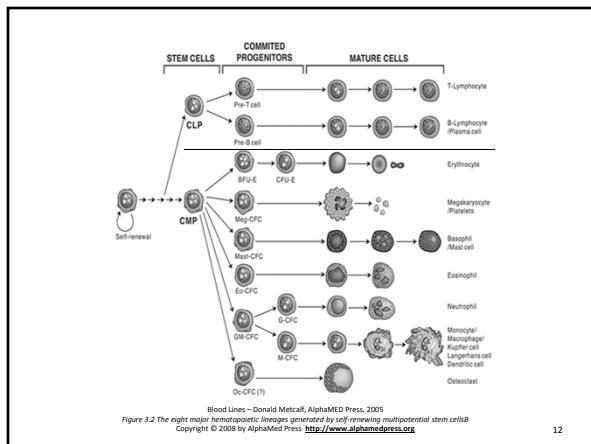Regenerative Medicine, 2006. http://www.dentalarticles.com/images/hematopoiesis.png

10

Proliferation and Differentiation

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

11

Blood Lines – Donald Metcalf, AlphaMED Press, 2005
Figure 3.2 The eight major hematopoietic lineages generated by self-renewing multipotential stem cellsB
Copyright © 2008 by AlphaMed Press http://www.alphamedpress.org

12

T-cell Differentiation

13

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
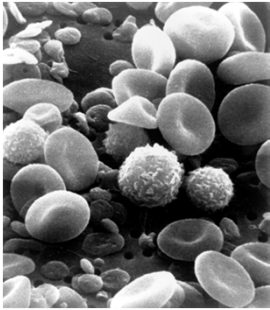
14

B-cell Differentiation

15

General Terms

- There are three major types of lymphocytes.
  - T-Cells
  - B-Cells
  - Natural Killer (NK) Cells

16

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.

17

Leukemia vs. Lymphoma

- Leukemia
  - Most commonly presents in the bone marrow or blood
  - When only the bone marrow is involved the histology is usually a leukemia
- Lymphoma
  - Most commonly presents in the lymphatic system

18

Lymphatic System

- Lymph nodes
  - Armpit: axillary nodes
  - Neck: cervical nodes
  - Groin: inguinal nodes
  - Intestine
    - Para-aortic nodes
    - Retroperitoneal nodes
- See appendix C of the Hematopoietic Manual

Lymph Node Structure

Afferent lymphatic vessel

Sinus

Capsule

Nodule

Valve to prevent backflow

Hilum

Efferent lymphatic vessel
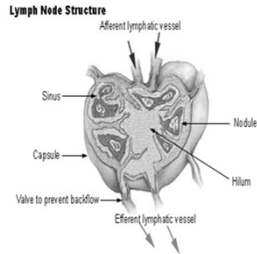
Image source: SEER Training Website

19

NAACCR

Lymphatic Structures

- Lymphatic organs
  - Waldeyer's ring
    - Pharyngeal tonsils (adenoids)
    - Palatine tonsils
    - Lingual tonsils

Adenoids

Lingual tonsils   Palatine tonsils

20

NAACCR
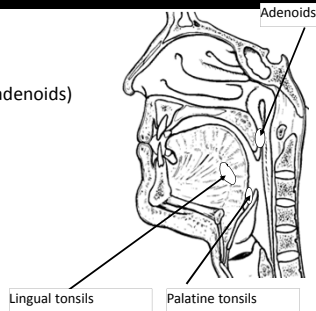
Lymphatic System

- Lymphatic organs
  - Spleen
  - Thymus
  - Peyer patches
  - Lymphoid nodules of the appendix

21

NAACCR

Extralymphatic vs. Extranodal

Lymph Nodes

Extranodal Lymphatic Sites

Extra Lymphatic (E)

Lymphatic System
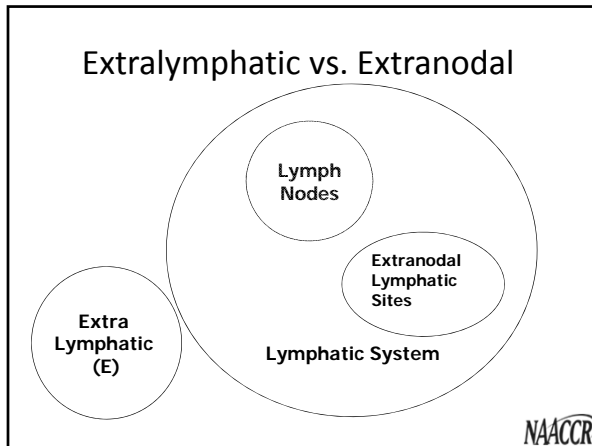
NAACCR

Diagnostic Process for Lymphoma

- Biopsy of most accessible lymph node
  - Not necessarily the primary site
- Bone marrow biopsy
  - Diagnostic purposes
  - Staging Purposes
- Additional testing
  - Immunophenotyping
  - Genetic testing

23

NAACCR

Diagnostic Process for Leukemia

1. Symptoms
   - Weight Loss
   - Weakness
   - Chronic fatigue
   - Bruising
2. CBC and/or peripheral blood smear
3. Bone marrow biopsy
4. More testing
   - Immunophenotyping
   - Genetic testing

24

NAACCR

HEMATOPOIETIC AND LYMPHOID DATABASE AND MANUAL

25   NAACCR

_____

_____

_____

_____

_____

_____

_____

Hematopoietic and Lymphoid Database and Manual

- Determine multiple primaries
- Assign primary site
- Assign histology
- Assign grade

26   NAACCR

_____

_____

_____

_____

_____

_____

_____

Hematopoietic and Lymphoid Database and Manual

- 2012 Hematopoietic and Lymphoid Database and Manual
  - Cases diagnosed 1/1/2012 and after
  - http://seer.cancer.gov/seertools/hemelymph
- 2010 Hematopoietic and Lymphoid Database and Manual
  - Cases diagnosed 1/1/2010 thru 12/31/2011
- ICD O 3 Hematopoietic Primaries Table
  - Cases diagnosed 1/1/2001 thru 12/31/2009
  - http://seer.cancer.gov/icd-o-3/

27   NAACCR

_____

_____

_____

_____

_____

_____

_____

Pop Quiz

- Patient 1
  - History of polycythemia vera (9950/3) diagnosed in 2009.
  - 1/12/12 he is diagnosed with Acute Myeloid Leukemia (9861/3)

Which rules do we use to determine if these are multiple primaries?

2012 Hematopoietic and Lymphoid Database and Manual

28

NAACCR

Using the Hematopoietic Database and Manual to Determine Multiple Primaries

1. Assign a code to each histology
   - (may also need to assign a primary site in some circumstances)
2. Use the Multiple Primary Manual to determine if any of the M rules apply
3. If rules M1-M15 do not apply use the Hematopoietic Database Calculator to determine if a single or multiple primaries

29

NAACCR

Example

- 1/1/12-A patient had a CT guided core biopsy of a pelvic mass positive for B-cell Non Hodgkin Lymphoma. He also had a biopsy of mediastinal lymph nodes positive for follicular lymphoma grade 1 of 2.

30

NAACCR

Example

- Step 1-Assign a code to each histology
  - B-Cell Non-Hodgkin Lymphoma
    - Non-Hodgkin Lymphoma, NOS (9591/3)
  - Follicular Lymphoma Grade 1(9695/3)
- Step 2-Review the multiple primary rules.
  - Rule M9 may apply
- Step 3-Use the Multiple Primary Calculator to confirm this is a single primary

31   NAACCR

Pop Quiz-Multiple Primaries?

- Patient 1
  - Follicular Lymphoma (9695/3) diagnosed in 2008
  - Diffuse large B-cell lymphoma (9680/3) diagnosed in 2010
    - Multiple Primaries? Yes/ No
- Patient 2
  - 1/13/12-A patient has an axillary lymph node dissection and is found to have two lymph nodes with Diffuse large B-cell lymphoma (9680/3) and four lymph nodes with Follicular Lymphoma (9695/3)
    - Multiple Primaries? Yes/ No

32   NAACCR

Transformation

- A chronic neoplasm is a neoplasm that can transform to an acute/more severe neoplasm
  - Follicular Lymphoma (9695/3) diagnosed in 2008
  - Diffuse large B-cell lymphoma (9680/3) diagnosed in 2010

Transformations

9680/3 Diffuse large B-cell lymphoma (DLBCL)

Same Primaries

9590, 9591, 9690, 9691, 9698

33   NAACCR

Diagnostic Work-up

- During the diagnostic workup the physician may start with a non-specific diagnosis (NOS) and as testing is completed, a more specific histology may be identified.
  - These diagnoses are **not** multiple primaries

34

NAACCR

Example

- A patient has a peripheral blood smear and CBC that indicate acute myeloid leukemia (9860/3). Additional samples are sent for genetic studies and immunophenotyping. Based on the results of these studies the physician states the patient has acute myeloid leukemia with inv(16)(p13.1q22) (9871/3).

35

NAACCR

Example

- Not necessary to code each histology or go through the multiple primary rules.
  - Acute myeloid leukemia was the provisional diagnosis
  - Acute myeloid leukemia with inv(16)(p13.1q22) was the complete diagnosis

36

NAACCR

You know you will have to refer to the Multiple Primary rules if…

- A sarcoma is diagnosed with or after a leukemia (M3)
- You are looking at more than one type of myelodysplastic syndrome (M4)
  - Refer to Table B4 in the Appendix for a list of myelodysplastic syndromes.
- You are looking at a follicular lymphoma and diffuse large b-cell lymphoma (M5)

37

NAACCR

You know you will have to refer to the Multiple Primary rules if…

- You are looking at a Hodgkin and non-Hodgkin lymphoma (M6-M8)
  - See table B10 in the appendix for a list of Hodgkin lymphoma's
- A more specific histology after an NOS histology (M9)
- You are looking at a chronic and acute phase of the same disease (M10-M15)

38

NAACCR

Coding Primary Site

- Always use the Heme DB when coding primary site
  - When a site code is listed in the primary site box, that is the only primary site that can be coded for that histology
  - Read the Abstractors Notes (when the Site Code Box says "See Abstractor Notes"). The Abstractor Notes contain the most common and also the rare primary sites for the histology.

39

NAACCR

Coding Primary Site

Disease Information

Name

B lymphoblastic *leukemia*/lymphoma, NOS

ICD-O-3 Code	Reportability	Primary Site(s)
9811/3	[REPORTABLE]	N/A - See Abstractor Notes and Module 7

Grade

6 - B-cell

Module Rule

Module 3: PH9, PH10

40

NAACCR

Example

- 1/1/12-A patient has a biopsy of a lymph node in the groin that is positive for chronic lymphocytic leukemia/small lymphocytic lymphoma. Imaging negative for any additional adenopathy. Bone marrow biopsy was negative.
  - Histology
    - 9823/3
  - Primary Site
    - C77.4
  - Grade
    - B-cell

41

NAACCR

Coding Primary Site for Lymphoma-Module 7

- Code to the specific lymph node region when multiple lymph node chains in the same region are involved (PH29)
  - See Appendix C for Lymph Node Reference Table
- Example:
  - Lymphoma is confined to the left and right axillary nodes.
    - Code C77.3 Lymph nodes of the axilla or arms

42

NAACCR

Coding Primary Site for Lymphoma

- Code the primary site as multiple lymph node regions, NOS C77.8 when multiple lymph node regions as defined by ICD-O-3 are involved and it is not possible to identify the lymph node region where the lymphoma originated (PH30).
  - CT showed lymphoma confined to the axillary and mediastinal lymph nodes.
    - Code C77.8 Lymph Nodes of Multiple Regions

43   NAACCR

Coding Primary Site for Lymphoma

- Code the primary site to C77.9 when multiple lymph nodes are present and the primary site cannot be identified (PH31).
  - Example: CT showed lymphoma throughout the neck, chest, abdomen.
    - Code C77.9 Lymph Nodes NOS

44   NAACCR

Coding Primary Site for Lymphoma

- Code to bone marrow (C42.1)if lymphoma is only present in the bone marrow (PH32).
- If the lymphoma is confined to an organ, code that organ as the primary site (PH33).
- If lymphoma originates in lymph nodes and extends into an organ, code to the lymph nodes (PH34).
- If lymphoma originates in an organ and involves that organs regional lymph nodes, code the organ of origin (PH36).

45   NAACCR

Coding Primary Site for Lymphoma

- Code the primary site to lymph nodes, NOS C77.9 when lymphoma is present in an organ and lymph nodes that are not regional for that organ and the origin cannot be determined even after consulting the physician (PH36).
- Code primary site to unknown primary site C80.9 when there is no evidence of lymphoma in lymph nodes AND the physician documents in the medical record that he/she suspects that the lymphoma originates in an organ(s) (PH37).

46   NAACCR

Question

- Patient with known mantle cell lymphoma in history on path report. Bone marrow biopsy read at my facility without involvement of bone marrow. There is no information concerning where lymphoma originated. Would the primary site be coded to C809 or C779?

47   NAACCR

Answer

- Code primary site lymph nodes, NOS C77.9.
  - See the Hematopoietic Manual, Module 7: Primary Site Rules for Lymphomas PH31.
  - The only time unknown primary site C80.9 is used is when there is no evidence of lymphoma in lymph nodes AND the physician documents that the lymphoma originates in an organ(s).

SEER SINQ 20120036
http://seer.cancer.gov/seerinquiry/index.php?page=view&id=20120036&type=q

48   NAACCR

Coding Histology

- Code the histology from definitive diagnostic method(s) (See Heme DB). Definitive diagnostic method(s) may be any of the following
  - Clinical diagnosis
  - Genetic test
  - Immunophenotyping
  - Cytology
  - Pathology
    - Final diagnosis
    - Comment on final diagnosis
    - Addenda to final diagnosis
    - CAP protocol

49

NAACCR

Ambiguous Terminology

- A list of ambiguous terms used to determine ***reportability*** is in the 2012 Hematopoietic Manual (page 14)

- Do ***not*** use ambiguous terms to code a specific histology.
  - Myeloproliferative disease, *probably* polycythemia vera
    - Code as Myeloproliferative disease, nos

50

NAACCR

Grade

- GRADE/DIFFERENTIATION

Code	Label
5	T cell; T-precursor
6	B cell; pre-B; B-precursor
7	Null cell; non T-non B
8	NK (natural killer) cell (effective with diagnosis 1/1/95 and after)
9	Cell type not determined, not stated or not applicable

51

NAACCR

Grade

- Do **not** use Table 13 on pages 16-17 of ICD-O-3 to determine grade.
- The only valid grade codes for hematopoietic neoplasms are 5, 6, 7, 8, AND 9
- Do **not** code based on descriptions "low grade," "intermediate grade," or "high grade"

52

NAACCR

Diagnostic Confirmation

- Records the best method of diagnostic confirmation of the cancer being reported at any time in the patient's history.
- Codes differ for solid tumors and hematopoietic primaries.
  - There is no priority hierarchy for coding *Diagnostic Confirmation* for hematopoietic and lymphoid tumors.

53

NAACCR

Diagnostic Confirmation

Code	Label	Definition
1	**Positive histology**	**Histologic confirmation (tissue microscopically examined).**
2	Positive cytology	Cytologic confirmation (no tissue microscopically examined; fluid cells microscopically examined).
3	**Positive histology PLUS** • **Positive immunophenotyping AND/OR** • **Positive genetic studies**	**Histology is positive for cancer, and there are also positive immunophenotyping and/or genetic test results.**

54

Diagnostic Confirmation

- For leukemia only, code 1 when the diagnosis is based only on the complete blood count (CBC), white blood count (WBC) or peripheral blood smear.
  - Do not use code 1 if the diagnosis was based on immunophenotyping or genetic testing using tissue, bone marrow, or blood.

55

NAACCR

Example

- Patient has peripheral blood smear on 1/1/12 and is found to have acute myelomonocytic leukemia (AML)
- Bone marrow is done on 1/15/12 that confirms AML
  - How do we code diagnostic confirmation?

56

NAACCR

Example

Abstractor Note

The peripheral blood (PB) and bone marrow (BM) have more than 20% blasts, neutrophils and their precursors, plus monocytes and their precursors. The monocyte component may be more evident in the PB than in the BM. The preliminary diagnosis may include AML with maturation, AML without maturation and AML. The diagnosis of AML is proven by BM biopsy. Patients typically present with anemia, thrombocytopenia, fever and fatigue. The WBC may be high.

Definitive Diagnostic Methods

Bone marrow ⟵

Peripheral blood ⟵ **Both tests fall under code 1 positive histology**

Immunophenotyping

57

NAACCR

Diagnostic Confirmation

- Code 3 is used when
  - The bone marrow, tissue, or blood (for leukemia) were suspicious for a neoplasm or provided a provisional diagnosis of a hematopoietic or lymphoid neoplasm

  AND

  - Genetic testing, immunophenotyping, or JAK2 EITHER
    - Confirmed the disease

    OR

    - Identified a more specific histology

58

NAACCR

Example

- A patient has a peripheral blood smear positive for acute myeloid leukemia
- Immunophenotyping is performed and indicates acute basophilic leukemia

59

NAACCR

Example

Definition

An acute myeloid leukemia in which the primary differentiation is to basophils.

Rare form of Acute myeloid leukemia where blasts are accompanied by abnormal basophils in all stages of differentiation.

Abstractor Note

Cutaneous involvement, organomegaly, lytic lesion, and symptoms relating to hyperhistaminemia may be present. Basophilic leukemia is differentiated from other AML subtypes with basophilia using basophilia immunological markers, light microscopic cytochemistry for myeloperoxidase and electron microscopy.

Definitive Diagnostic Methods

Bone marrow
Peripheral blood ⟵ **Histologic and immunophenotyping**
Immunohistochemistry ⟵ **Use code 3**

60

NAACCR

Diagnostic Confirmation

- Do not use code 3 when the genetic testing and/or immunophenotyping are used to rule out other diseases. In this case, the impression on the report will be such as "rule out CML."
  - Do not use code 3 when the genetics and/or immunophenotyping are not positive for the neoplasm.

61

NAACCR

Diagnostic Confirmation

Code	Label	Definition
4	Positive microscopic confirmation, method not specified	Microscopic confirmation is all that is known. It is unknown if the cells were from histology or cytology
5	Positive laboratory test/marker study	A clinical diagnosis of cancer is based on laboratory tests/marker studies which are clinically diagnostic for cancer.
6	Direct visualization without microscopic confirmation	The tumor was visualized during a surgical or endoscopic procedure only with no tissue resected for microscopic examination.

62

Diagnostic Confirmation

Code	Label	Definition
7	Radiography and other imaging techniques without microscopic confirmation	The malignancy was reported by the physician from an imaging technique report only.
8	**8 Clinical diagnosis only, other than 5, 6 or 7**	**The malignancy was reported by the physician in the medical record.**
9	Unknown whether or not microscopically confirmed	A statement of malignancy was reported in the medical record, but there is no statement of how the cancer was diagnosed (usually nonanalytic).

63

Diagnostic Confirmation

- Assign code 8 when the case was diagnosed by any clinical method that can not be coded as 6 or 7.
  - A number of hematopoietic and lymphoid neoplasms are diagnosed by tests of exclusion where the tests for the disease are equivocal and the physician makes a clinical diagnosis based on the information from the equivocal tests and the patient's clinical presentation.

64

NAACCR

Question

- Could you give me step by step instruction of how to code the diagnostic confirmation for thrombocythemia? Do we use the Hematopoietic Manual Rules?

65

NAACCR

Diagnostic Confirmation

Abstractor Note

Disease is usually suspected when the patient has a CBC or peripheral blood smear that shows an overproduction of platelets. The diagnosis is clinical (diagnosis of exclusion); the physician diagnoses this disease by excluding other myeloproliferative neoplasms and correlating those equivocal results with the patient's symptoms. Bone marrow and blood are the principle sites of involvement. The spleen does not show significant extramedullary hematopoiesis but is a sequestration site for platelets. In some cases this disorder may be progressive, and only rarely evolves into acute myeloid leukemia or myelofibrosis.

50-60% of patients will have a positive JAK2.

Definitive Diagnostic Methods

Clinical diagnosis

Genetics

66

JAK2

Answer

- Code diagnostic confirmation as 8, clinical diagnosis
- Thrombocythemia is a diagnosis of exclusion
  - Elevated platelet count could indicate a number of diseases
    - Polycythemia vera (PV)
    - Chronic myelogenous leukemia (CML)
    - Idiopathic myelofibrosis
    - Myelodysplastic syndrome (MDS)

67

NAACCR

Reportable Cases

- If a patient receives treatment at your facility, report the case even if the diagnostic tests are inconclusive, equivocal, or negative
- If the diagnostic tests are inconclusive, equivocal, or negative but the physician gives a positive clinical diagnosis, report the case

68

NAACCR

Practice Case 1

- A patient is diagnosed with a diffuse large B-cell lymphoma originating in the stomach in 2004 at another facility. The patient had a recurrence in the inguinal lymph nodes in 2012.

69

NAACCR

Practice Case 1

- How many primaries does this patient have?

- What is the histology for each primary?

- What is the primary site for each primary

- What is the Grade for each primary?

70   NAACCR

Practice Case 2

- 1/15/12 TAH/BSO final pathologic diagnosis:
  - 2 cm adenocarcinoma of the endometrium confined to endometrium
  - Diffuse large B-cell lymphoma in three parametrial lymph nodes and focal involvement by follicular lymphoma in a fourth lymph node.
  - Further diagnostic work-up did not reveal any additional disease.

71   NAACCR

Practice Case 2

- How many primaries does this patient have?

- What is the histology for each primary?

- What is the primary site for each primary

- What is the Grade for each primary?

72   NAACCR

Practice Case 3

- 1/1/12 patient had a peripheral blood smear that is positive for chronic myelomonocytic leukemia
- 1/20/12 A bone marrow biopsy was done that showed acute myeloid leukemia.
- 1/23/12 A sample was sent for cytochemical testing. This test indicated Acute promyelocytic leukemia (AML with t(15;17)(q22;q12)) PML/RARA

73

NAACCR

Practice Case 3

- How many primaries does this patient have?

- What is the histology for each primary?

- What is the primary site for each primary

- What is the Grade for each primary?

74

NAACCR

Practice Case 4

- 1/1/12-Patient presents with right cervical adenopathy. A biopsy of a cervical node indicate small lymphocytic leukemia. A bone marrow biopsy was diagnostic of chronic lymphocytic leukemia/small lymphocytic lymphoma

75

NAACCR

Practice Case 4

a. How many primaries does this patient have?

b. What is the histology for each primary?

c. What is the primary site for each primary

d. What is the Grade for each primary?

76   NAACCR

Practice Case 5

- 1/1/12-Patient had a biopsy of a cervical lymph node. The pathology report indicates Hodgkin lymphoma, nodular sclerosis, grade 2 and large B-cell lymphoma, diffuse.

77   NAACCR

Practice Case 5

a. How many primaries does this patient have?

b. What is the histology for each primary?

c. What is the primary site for each primary

d. What is the Grade for each primary?

78   NAACCR

STAGING

79

NAACCR

Site Definitions for Lymphoma

- Lymphatic sites
  - Lymph nodes (C77._)
  - Thymus (C37.9)
  - Spleen (C42.2)
  - Waldeyer's ring
  - Peyer's patches
  - Lymphoid nodules of appendix
- Extralymphatic sites
- Diaphragm

80

NAACCR

Anatomic Stage/Prognostic Groups
Hodgkin & Non-Hodgkin Lymphoma

- Stage I
  - Involvement of single lymphatic site OR single extralymphatic organ
- Stage II
  - Involvement of 2 or more lymph node regions on same side of diaphragm OR single extralymphatic organ with regional node involvement
- Stage III
  - Involvement of lymph node regions on both side of diaphragm
- Stage IV
  - Disseminated involvement of 1 or more extralymphatic organs

81

NAACCR

Anatomic Stage/Prognostic Groups
Hodgkin & Non-Hodgkin Lymphoma

- E = Involvement of extralymphatic organ
- S = Involvement of spleen

82

NAACCR

CS Extension:
Hodgkin & Non-Hodgkin Lymphoma

- Stage I CS Extension codes
  – Code 100: Single lymph node region (I)
  – Code 110: Single extralymphatic organ/site (IE)
  – Code 120: Spleen only (IS)
    - Primary site must be spleen (C42.2)

83

NAACCR

CS Extension:
Hodgkin & Non-Hodgkin Lymphoma

- Stage II CS Extension codes
  – Code 200: 2 or more lymph node regions on the SAME side of the diaphragm (II)
  – Code 210: Single extralymphatic organ/site (IIE)
    - WITH involvement of its regional lymph node(s)
    - Direct extension to adjacent organs or tissues
  – Code 220: Spleen PLUS lymph node(s) BELOW the diaphragm (IIS)
  – Code 230: Spleen PLUS involvement of a single extralymphatic organ/site BELOW the diaphragm (IIES)

84

NAACCR

CS Extension:
Hodgkin & Non-Hodgkin Lymphoma

- Stage III CS Extension codes
  - Code 300: Lymph node regions on both sides of the diaphragm (III)
  - Code 310: Extralymphatic organ/site PLUS involvement of lymph node(s) on OPPOSITE side of diaphragm (IIIE)
  - Code 320: Spleen PLUS lymph node(s) ABOVE the diaphragm OR PLUS lymph nodes on both sides of the diaphragm (IIIS)
  - Code 330: 320 + 310 (IIIES)

85

NAACCR

CS Extension:
Hodgkin & Non-Hodgkin Lymphoma

- Stage IV CS Extension code
  - Code 800
    - Diffuse or disseminated (multifocal) involvement of one or more extralymphatic sites with or without lymph node involvement
    - Involvement of isolated extralymphatic organ in absence of involvement of adjacent lymph nodes, but in conjunction with disease in distant sites
    - Metastasis to bone marrow, liver, lung

86

NAACCR

CS Extension Summary Table
Hodgkin & Non-Hodgkin Lymphoma

Nodal/ Lymphatic	Extranodal/ Extralymphatic	Spleen (involvement)
100	110	120
200	210	220, 230
300	310	320, 330
800	800	800
999	999	999

87

NAACCR

Pop Quiz

- Final pathologic diagnosis is large b-cell lymphoma of the thymus. No other lymphadenopathy or organ enlargement per CT scan. What is the code for CS Extension?
  - 100: Involvement of a single lymph node region
  - 110: Localized involvement of a single extralymphatic organ/site in the absence of any lymph node involvement
  - 800: Diffuse or disseminated (multifocal) involvement of ONE OR MORE extralymphatic organ(s)/site(s) WITH OR WITHOUT associated lymph node involvement
  - 999: Unknown

88   NAACCR

CS Tumor Size Ext/Eval

Code	Description
0	Clinical evidence
2	Autopsy (tumor suspected or diagnosed prior to autopsy)
3	Staging laparotomy done
8	Autopsy (tumor unsuspected or undiagnosed prior to autopsy)
9	Unknown

89   NAACCR

SSF1: Associated with HIV/AIDS

Code	Description
000	Not associated with HIV/AIDS
010	Associated with HIV/AIDS
988	Not applicable: Information not collected for this case
999	Unknown

- **Use code 999 when no documentation about HIV/AIDS status**

90   NAACCR

SSF2: Suspected Symptoms at Diagnosis

Code	Description
000	No B symptoms (asymptomatic)
010	Any B symptoms: Night sweats, unexplained fever, unexplained weight loss
020	Pruritus (recurrent and unexplained)
030	020 + 010
988	Not applicable: Information not collected for this case
999	Unknown

91

NAACCR

Prognostic Scoring System

- SSF3: International Prognostic Index (IPI)
  - Tracks prognostic variables for non-Hodgkin lymphoma
- SSF4: Follicular Lymphoma Prognostic Index (FLIPI)
  - Tracks prognostic variables for follicular non-Hodgkin lymphoma
- SSF5: International Prognostic Score (IPS)
  - Tracks prognostic factors for Hodgkin lymphoma

92

NAACCR

Prognostic Scoring System

- Code named score in appropriate SSF If index or score named and point value given; code other two SSFs as 999
- If index or scoring system not named
  - Code all 3 SSFs as 999 if point value is 5 or less
  - Code the point value in SSF5 if score is 6 or 7 and code SSF3 and SSF4 as 999
  - Code all 3 SSFs as 999 if risk stated as "low," intermediate," or "high"
- Point value takes priority over the risk category if both are stated and the score is named
- Code only the statement/score/index documented by clinician

93

NAACCR

Pop Quiz

- Cerebral diffuse large B-cell lymphoma. Physical exam showed no lymphadenopathy, no organ enlargement, and no other abnormalities. What is the code for SSF1 (Associated with HIV/AIDS)?
  - 000: Not associated with HIV/AIDS
  - 010: Associated with HIV/AIDS
  - 999: Unknown

94

CS Extension: HemeRetic Neoplasms

- Code 100: Localized disease
  - (Single/solitary/unifocal/isolated):
    - May be coded for: Mast cell sarcoma (9740); malignant histiocytosis (9750); Langerhans cell histiocytosis (9751); histiocytic sarcoma (9755); Langerhans cell sarcoma (9756); dendritic cell sarcoma (9757, 9758); myeloid sarcoma (9930)
- Code 800: Systemic disease
  - Always use for leukemia
    - *Except* death certificate only case diagnosed with leukemia
- Code 999: Unknown

95

SSF1: JAK2

Code	Description
000	JAK-2 result stated as negative
010	JAK2 positive for mutation V617F in exon 14
020	JAK2 positive for mutation of exon 12
800	JAK2 positive for other specified mutation
810	JAK2 positive for more than one mutation
850	JAK2 positive NOS; specific mutation(s) not stated
988	Not applicable: Information not collected for this case
997	Test ordered, results not in chart
998	Test not done
999	Unknown

96

CS Extension: Plasma Cell Disorders Including Myeloma

Code	Description
110	Single plasmacytoma lesion WITHOUT soft tissue extension or unknown if soft tissue extension (9731)
200	Single plasmacytoma lesion WITH soft tissue extension (9731)
300	Single plasmacytoma lesion occurring in tissue other than bone (9734)
400	Multiple osseous or multiple extraosseous plasmacytoma lesions (9731, 9734)

97

NAACCR

CS Extension: Plasma Cell Disorders Including Myeloma

Code	Description
500	Plasmacytoma, NOS (9731) Not stated if single or multiple, not stated if osseous or extraosseous
810	Plasma cell myeloma/multiple myeloma/myelomatosis (9732)
820	Myeloma, NOS Excludes plasma cell myeloma or multiple myeloma (see code 810)
999	Unknown

98

NAACCR

CS Lymph Nodes: Plasma Cell Disorders Including Myeloma

Code	Description
000	For 9734 only: No regional lymph node involvement
100	For 9734 only: Regional lymph node metastasis
987	Not applicable: Use for codes 9731 & 9732
988	Not applicable: Information not collected for this schema
999	Unknown

99

NAACCR

SSF2: Durie-Salmon Staging System

Code	Description
010	Durie Salmon Stage IA
020	Durie Salmon Stage IB
030	Durie Salmon Stage I NOS
040	Durie Salmon Stage IIA
050	Durie Salmon Stage IIB
060	Durie Salmon Stage II NOS
070	Durie Salmon Stage IIIA
080	Durie Salmon Stage IIIB
090	Durie Salmon Stage III NOS
987	Not applicable: Use for codes 9731 & 9734
988	Not applicable: Information not collected for this case
999	Unknown

100

SSF3: Multiple Myeloma Terminology

Code	Description
000	Multiple myeloma/Plasma cell myeloma with no other modifiers Multiple myeloma, NOS; Myeloma, NOS
010	Asymptomatic myeloma
020	Early or evolving myeloma
030	Inactive, indolent, or smoldering myeloma
080	Other terminology describing myeloma
100	Any combination of terms in codes 010-080
987	Not applicable: Use for codes 9731 & 9734
988	Not applicable: Information not collected for this case
999	Unknown

101

Pop Quiz

- 3/1/2012: Based on CT scan and lab work, plasmacytoma of right rib.
- 3/15/2012: Bone marrow biopsy diagnoses multiple myeloma.
- What is the code for CS Extension?
  - 110: Single plasmacytoma lesion WITHOUT soft tissue extension or unknown if soft tissue extension
  - 200: Single plasmacytoma lesion WITH soft tissue extension
  - 300: Single plasmacytoma lesion occurring in tissue other than bone
  - 810: Plasma cell myeloma/multiple myeloma/myelomatosis

102

TREATMENT

103

NAACCR

Surgery

- 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

104

NAACCR

Example

- A patient presents with bilateral cervical adenopathy. Two lymph nodes are surgically excised from the right side of the neck. These lymph nodes are positive for B-cell lymphoma.
  - Excisional biopsy of the two lymph nodes would be coded as 02-Diagnostic Staging Procedure
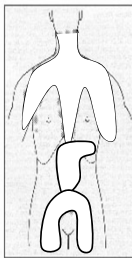
105

NAACCR

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

106

NAACCR

Radiation Therapy
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
- Involved Field Radiation Therapy (IFRT)

Image source: SEER Training Website

NAACCR

Radiation Therapy
Other Hematopoietic Diseases

- Radiation is not usually part of treatment for leukemia
- Patients with other hematopoietic diseases may receive radiation therapy
  - Langerhan cell histiocytosis
  - Histiocytic sarcoma
  - Waldenstrom macroglobulinemia

108

NAACCR

Radiation Therapy for Lymphoma

- Assign code 33, whole body in data item, regional treatment modality
  - Drugs combine the cell targeting ability of a monoclonal antibody with the additional cell killing ability of a radioactive particle, or radioisotope
    - Bexxar (iodine-131 tositumomab)
    - Zevalin (yttrium 90 ibritumomab tiuxitan)

109   NAACCR

Systemic Therapy

- Primary treatment for most hematopoietic and lymphoid neoplasms
  - Varies widely based on diagnosis
- Use SEER Rx to determine if systemic treatment is chemotherapy, hormone therapy, immunotherapy, or other therapy.
  - http://seer.cancer.gov/tools/seerrx/

110   NAACCR

Systemic Therapies

- Rituximab
- CHOP
- ABVD
- Anthracycline

111   NAACCR

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)

112

NAACCR

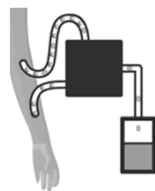## Immunotherapy

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

113

NAACCR

Hematologic Transplant

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

114

Image source: http://www.lymphoma-net.org/transplantation.cfm

NAACCR

Other Treatment

- Aspirin
  - Code as 'Other' therapy for essential thrombocythemia ONLY otherwise it is a differentiating agent (do not code)
- Anagrelide HCl
  - Code as 'Other' therapy for essential thrombocythemia ONLY otherwise it is a differentiating agent (do not code)

115   NAACCR

QUESTIONS?

116   NAACCR

Coming up!

- 9/6/12
  - Coding Pitfalls
- Register now for the 2012-2013 Cancer Registry & Surveillance Webinar Series
  - http://www.naaccr.org/EducationandTraining/Webinars.aspx

And the winners of the
117   **fabulous prizes are….**   NAACCR