In this presentation, we will discuss the rules for abstracting leukemia and other hematopoietic diseases. You should follow along in your ICD-O-3 and CS manuals. Again, this presentation will review 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
This presentation contains 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 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.
Listed on the slide are the code ranges that are included in this presentation. It is important to note that new codes went into effect for cases diagnosed on or after 1/1/2010. These new codes are NOT in the ICD-O-3 manual.

The first few pages of the Hematopoietic CS schema, which lists all the applicable histology codes for this schema, has been updated to include all the newly reportable codes that are staged under the Hematopoietic schema. These new codes are also included in the 2010 Hematopoietic and Lymphoid neoplasm rules (Table D1a: New Histology Terms and Codes – Alphabetic List, and Table F: Master Code Lists). The diseases with these new codes will follow the same staging rules discussed in this presentation.
Leukemia can be grouped 2 different ways.

There are 2 by the type of white blood cell that is affected – lymphoid and myeloid. There are 2 by how quickly the disease develops and gets worse – chronic and acute.

As a result, there are 4 groups: CLL, CML, ALL, AML.
### Four main types of Leukemia

<table>
<thead>
<tr>
<th>Type</th>
<th>Characteristics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute lymphocytic leukemia (ALL)</td>
<td>Affects children and adults&lt;br&gt;More common among children&lt;br&gt;Slightly more than half of all childhood leukemias</td>
</tr>
<tr>
<td>Acute myelogenous leukemia (AML)</td>
<td>Affects children and adults&lt;br&gt;Just under half of all childhood leukemias</td>
</tr>
<tr>
<td>Chronic lymphocytic leukemia (CLL)</td>
<td>Affects adults&lt;br&gt;Almost twice as common as CLL</td>
</tr>
<tr>
<td>Chronic myelogenous leukemia (CML)</td>
<td>Affects mostly adults&lt;br&gt;Very rare in children</td>
</tr>
</tbody>
</table>

Here are some common characteristics of the four main types of leukemia. While both children and adults can develop leukemia, certain types are more common in one age group than in another. ALL is more common in children. On the other end of the spectrum, CML is more common in adults.
Diagnostic Workup

Often found during workup for an unrelated health problem

General symptoms:
- Weight loss, bruising, bleeding, anemia, fatigue, bone pain

Lab work:
- Blood cell counts and blood tests
  - Lymphocytosis – too many white cells
- Bone marrow aspirations and biopsies
- Lumbar puncture

Because of the way leukemia is diagnosed and treated – usually in the doctor’s office – you have to be really careful that you are not missing cases. Most of the time you are going to pick them up if they come in for treatment. But, you should make sure you are not missing cases through other types of visits. IE: if the lab work is done in your facility. You can’t always count on having a pathology report. Because leukemia does not usually form tumors, imaging test are not always helpful and are usually only done to look for infections and other problems.
There is a complete list of histologies that are included in the hematopoietic staging scheme on the first few pages of the hematopoietic staging schema in the CS Manual. This list includes the new histology codes that became effective on January 1, 2010. Review this list carefully to ensure you are using the correct staging schema.
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.

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.
There are only TWO CS data fields that needs to be coded for hematopoietic primaries. The rest are either all 8’s, all 9’s, or 988. Notice that tumor size is 988, not 999.

For the CS Extension data item there are only 3 codes to choose from. Most cases will be coded to 800 – Systemic. All leukemias and anything multifocal such as multiple myeloma or systemic such as refractory anemia are always a code of 800.
There may be times when code 100 is appropriate. This code applies when the involvement is described as being a single lesion. There are 9 histologies listed under this code, one of which is solitary myeloma/plasmacytoma. This condition was discussed earlier in the Hematopoietic Rules educational presentations. If the plasmacytoma is solitary (there is only one lesion) then assign the CS Extension code of 100.

Review the histology codes included in code 100 carefully and make sure that the histology code for the case is listed. If the diseases listed for this code are “localized”, you may use code 100.

Note: Langerhans cell histiocytosis, code 9751/1, was changed to a behavior code of /3 effective 1/1/2010. This disease is included in the code 100 group.
Code 800 includes diseases from code 100 that have become systemic (or there are multiple lesions). All leukemia and myelodysplastic histologies should ALWAYS be coded 800. These are by nature systemic diseases.

For the hematopoietic diseases that became reportable with the 2010 Hematopoietic and Lymphoid neoplasms rules, code 800 is the appropriate code.

Only use code 999 for those codes that are listed in code 100 where you don’t know whether they are localized or systemic (or single or multiple lesions).
The data items listed on this slide are “not applicable” for Hematopoietic diseases. 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.
JAK2 is a gene mutation that increases susceptibility to several Myeloproliferative neoplasms (MPN). Testing for the JAK2 mutation is done on whole blood. Nearly all people with polycythemia vera and about half of those with essential thrombocytopenia and primary myelofibrosis have the mutation. The most common mutations used now are the V617F in exon 14 and exon 12.

When JAK2 is positive the MPN is definitely reportable; however, JAK2 itself does not specify which MPN is present. Other diagnostic tests are required to determine the specific histology. Note: you should use the Hematopoietic rules to make a final decision as to reportability.

Code the result of the JAK-2 test as documented in a laboratory test or elsewhere in the medical record. Code this field for any hematopoietic, reticuloendothelial, immunoproliferative, myeloproliferative, or myelodysplastic disease for which JAK-2 is tested.

If you have a JAK 2 positive with no other information, use code 850.

For those diseases where JAK 2 is not mentioned in the record, or it is a disease where it is normally not done (i.e. leukemia and multiple myeloma), code 999.
Clinical History:
Chronic Myelod Leukemia, Leukoeytosis

DIAGNOSIS:
A JAK2 AND BCR/ABL BY PCR QUANT. ANALYSIS WAS PERFORMED BY NEOGENOMICS LABORATORIES IN FORT MYERS, FLORIDA UNDER THEIR ACCESSION NO: . INTERPRETED BY . PATHOLOGIST AS FOLLOWS:

JAK2: NEGATIVE FOR JAK2 (V617F) MUTATION. THERE IS NO EVIDENCE OF THE JAK2 (V617F) POINT MUTATION BY REAL-TIME PCR ANALYSIS.

BCR/ABL: POSITIVE FOR BCR/ABL BY QUANTITATIVE PCR METHODOLOGY. BCR-ABL:ABL RATIO = 98.15%

This is a sample report showing JAK2 results.
Additional Comments related to the Hematopoietic Rules
Leukemia along with the other types of disorders and syndromes in the 9800-9992 range are a malignant disease of the bone marrow. Therefore the site code will almost ALWAYS be coded to C42.1.

Refer to the Hematopoietic Coding Rules manual for specific instructions on coding the primary site as there may be situations where this differs. However, if you have a histology in the 9800-9992 range, and the site is NOT C42.1, consider it a flag to double check to make sure you have followed the rules correctly and have assigned the correct site code.
If you look through the ICD-O-3 manual at the leukemia terms and codes, you may notice that some codes contain unusual terminology. The purpose of the next few slides is to describe the application of cytogenetics and FAB classification in the ICD-O-3 manual.

The first topic we will discuss is cytogenetics.

Because leukemia is a systemic disease that starts in the bone marrow, traditional staging is not useful. Instead, physicians look at the cellular level to identify not only the subtype, but also the prognosis, and the likely response to treatment. Clinicians use cytogenetic testing to identify certain types of myeloid leukemia. The results of the cytogenetic testing are written in shorthand form that describes which chromosome changes are present.

For example, for translocation, this means that part of chromosome 1 is now located on chromosome 2 or vice versa. An inversion means that part of chromosome 16 is upside down and is now in reverse order.
Look at pages 102-103 in your ICD-O. Notice beside selected myeloid leukemias, the histologic term is followed by a series of numbers written in parentheses.

For example, under Acute Promyelocytic Leukemia (9866/3), the following shorthand is listed: t(15;17)(q22;q11-12).

Based on this information, this means that part of chromosome 15 is now located on chromosome 17 and vice versa. And, part of chromosome q22 is now located on chromosome q11-12 and vice versa.

While it is not vital that you know that it means that exactly, it is important that you understand, in general, what it is referring to so when you do see one, you can code it correctly.

Under code 9866, notice that acute MYELOID leukemia is listed with the same shorthand. If the diagnosis doesn’t specify the shorthand or the FAB type, then code to Acute Myeloid Leukemia, NOS which is code 9861/3 – listed just above it.
This slide lists some of the commonly used genetic shorthand symbols.
The second area to bring to your attention is the FAB Classification.

**Refer to the handout provided in the course content called ‘Cytological Classification for Leukemia’**.

The most popular classification method for acute leukemia is the French-American-British (FAB) system. According to the FAB classification, AML is divided into eight subtypes and ALL is divided into three subtypes. Each subtype is described according to the morphology of the abnormal cell as you can see on the table in the handout.
Remember, the FAB system was for acute leukemias. Find acute leukemia in the ICD-O alphabetic section (p158).

Go down to the L's for lymphoblastic (under acute). Do you see any of the FAB L1-3 types? There is one - L2.
Now go on down to the M's for myeloid (still under acute). Do you see any of the FAB M0-7 types? There is one - M6.
Follow this down to where chronic leukemia ends and it starts with common, cortical, erythroid on pg 159.
Turn the page, after “e” comes “f”, at the top of page 160 we have FAB and under it all the types are listed.

The table on the handout cross-references the FAB subtype, the ICD-O-3 code and the terminology.
For example, look at M1. On page 160, M1 is 9873.
Go to 9873 in the numerical section – p 103. You can see the terminology that goes with that – Acute myeloid leukemia w/o maturation. This is what is on the table.
Go back to Acute myeloid in the alphabetic section – p158. Look for acute myeloid leukemia w/o maturation. M1 is not listed with it as a synonym.

So what if the diagnosis is M2 acute myelocytic leukemia? Even though it doesn’t say “with maturation” in the diagnosis, because we have a FAB classification of M2, we should code to 9874/3. The M2 is telling us the “with maturation” part.

If the FAB classification is not specified, you would have to use the NOS code of 9861/3. This is an excellent example of where looking something up in the alphabetic AND the numerical sections are an important concept.

AND, knowing that the M0-7 subtypes are part of the FAB system and knowing that these are listed under “FAB” in the ICD-O is important in seeing that you assign the correct and most specific code. Again, knowing exactly what M1 or M2 means is not as important as knowing how to code them when the term is included in the final diagnosis.
Disease transformation is an important part of the Hematopoietic rules.
Disease progression is defined as the worsening of a disease over time. It is most often used for chronic or incurable diseases.

As you learned in the staging timing rules, “disease progression” after the diagnosis is established is not included in calculating the stage. Disease progression applies to solid tumors as well. Disease progression in solid tumors usually involves further tumor extension, LN metastasis, body cavity metastases, and/or blood-borne metastases.

Hematopoietic disease can progress as well. Examples include:
• Smoldering multiple myeloma to “full blown” multiple myeloma
• Early stage/asymptomatic CLL to late stage/symptomatic CLL

For both solid tumor and hematopoietic disease progression, the characteristics are the same. The cell type is the same, the “function” is the same, and the genetics are the same. There is also a change in the symptoms, treatment, and life expectancy.
Transformation is a change in the nature, function, or condition of the disease. There is often a change in potential or cell type or the cell is undergoing genetic transformation. As with progression, there is also a change in the symptoms, treatment, and life expectancy.

Example: Polycythemia vera (PCV) transforming to acute myeloid leukemia (AML). For each of these the characteristics are different:

**Cell Origin**
PV: Hematopoietic stem cells
AML: Myeloid line of white blood cells

**Characteristics**
PV: Too many red blood cells causing blood to thicken
AML: 20% or more blasts in blood or BM

**Treatment**
PV: Phlebotomy, aspirin, sometimes Anagrelide
AML: Chemo, stem cell transplant
**Survival**
PV: Treated properly, virtually normal
AML: Develops quickly. Untreated, is fatal in a few months. Treated: 33% free of disease at 4 year interval (<65 Y/O)

**Conclusion**
Transformations are **NOT the equivalent of disease progression**. Transformations are considered to be a new primary as there is a different cell type, genetics, treatment, and survival. It is important to capture the incidence count for both chronic and acute. You should use the Hematopoietic Rules manual to make this determination of the number of primaries.
Sequence Number

- Takes into consideration ALL cancer primaries during the lifetime of the patient
- Only those neoplasms that were REPORTABLE AT THE TIME OF DIAGNOSIS are to be taken into consideration
- Changes in reporting rules:
  - ICD-O-2 → ICD-O-3 (01/01/2001)
  - Use the ICD-O-3 Appendices
  - Hematopoietic DB (01/01/2010)
  - Use the Hematopoietic Rules manual

The year in which the hematopoietic primary was diagnosed depends on whether or not it is reportable and should be factored into the sequence number. Don't forget about the changes when the ICD-O-3 came into effect in 2001 which added some hematopoietic primaries with a reportable behavior code of 3. And, the changes when the Hematopoietic DB came into effect in 2010 which added some new codes and some existing codes were changed from /1 to /3.

For example, refractory anemia was not reportable until the implementation of the ICD-O-3 in 2001. These changes can be found in the appendices in the back of your ICD-O-3 manual. This may not be specified in the Hematopoietic DB as the DB does not address assigning the sequence code. Therefore, you may need to use the ICD-O-3 appendices to determine when a particular condition became reportable.
Concluding remarks

- Remember which fields are not applicable (majority)
- When coding, double check “code 100” to see if your histology is applicable for this code
- Leukemias and MDS (9800-9989) are ALWAYS coded 800 in CS Extension
- SSF for JAK2 has been added

This concludes Part 2.
Please return to the course content.