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Correctly Applying Diagnostic Confirmation for Hematopoietic and Lymphoid Primaries

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During a review of the Diagnostic Confirmation field for hematopoietic and lymphoid primaries, we observed that many of us inadvertently fall into the trap of automatically using code 1 (positive histology) as we do for more than 90% of the solid tumors we accession annually. In order to improve the accuracy in assigning a code to this field, we need to consistently remember a few rules:

1. The codes and instructions for hematopoietic and lymphoid neoplasms are different from the codes for solid tumors.
2. While microscopic confirmation (codes 1-4) has priority order over clinical diagnosis only (codes 5-8), there is no priority order or hierarchy for coding Diagnostic Confirmation for hematopoietic and lymphoid neoplasms.
3. There is no time frame associated with coding this field.

The Diagnostic Confirmation field reflects the “best method” used to confirm the presence of cancer. It is an indicator of diagnostic precision. The best diagnostic method is the one that reliably gives the same result when a single sample is tested repeatedly. For solid tumors, the best method is considered a positive histology (Code 1). However, for hematopoietic and lymphoid primaries it is a combination of a positive histology PLUS positive immunophenotyping and/or genetic test results (Code 3).

As stated in rule 3 above, we need to keep in mind that the best method of confirmation could occur at any time throughout the entire course of the disease and is not limited to the confirmation at the time of initial diagnosis. For example, if we initially code the Diagnostic Confirmation to 1 to reflect the positive histology observed at diagnosis and later learn immunophenotyping was performed on the same or a different positive pathology specimen, we should update the Diagnostic Confirmation code to 3.

We need to understand the various codes to make sure we are correctly coding Diagnostic Confirmation for these cases. Start by referring to Table 1 for the Diagnostic Confirmation codes and definitions used for hematopoietic and lymphoid neoplasms before reading further to handle selected cases we’ve come across in the database.

Table 1
Diagnostic Confirmation
Codes, Heme Manual Descriptions and STORE Definitions

Microscopically Confirmed

Code	Heme Manual Descriptions	STORE Definitions
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: <ul style="list-style-type: none"> • Positive immunophenotyping AND/OR • Positive genetic studies (Effective for cases diagnosed 1/1/2010 and later only.)	Histology is positive for cancer, and there are also positive immunophenotyping and/or genetic test results. For example, bone marrow examination is positive for acute myeloid leukemia. (9861/3) Genetic testing shows AML with inv (16) (p13.1q22) (9871/3). Used only for hematopoietic and lymphoid neoplasms (M-9590/3-9992/3).
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.

Not Microscopically Confirmed

Code	Heme Manual Descriptions	STORE Definitions
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.
7	Radiology and other imaging techniques without microscopic confirmation	The malignancy was reported by the physician from an imaging technique report only.
8	Clinical diagnosis only (other than 5, 6 or 7)	The malignancy was reported by the physician in the medical record.

Confirmation Unknown

Code	Heme Manual Description	STORE Definition
9	Unknown whether or not microscopically confirmed; death certificate only	A statement of malignancy was reported in the medical record, but there is no statement of how the cancer was diagnosed (usually nonanalytic).

Source: Hematopoietic and Lymphoid Neoplasm Coding Manual

Source: STORE 2018 for Store Definitions

Using Diagnostic Confirmation Code 1 vs. Code 3

Generally, the **histology provides conditional diagnoses and the specific histologic type is determined through immunophenotyping or genetic testing**. Code 1 is used when the diagnosis is based on tissue from lymph node(s), organ(s), other tissues from biopsy, surgery, etc., or from a bone marrow aspiration, bone marrow biopsy or peripheral blood smear. In addition, for leukemia only (9800/3-9948/3), code 1, positive histology, includes a complete blood count (CBC) or white blood count (WBC). However, code 1 does not have priority over Code 3, which is used if a patient has a positive histology and the three following conditions are met:

- Genetic testing and/or immunophenotyping are described in the Hematopoietic Database "Definitive Diagnostic Methods" field; AND
- Genetic testing and/or immunophenotyping were done; AND
- Genetic testing and/or immunophenotyping confirms the diagnosis or identifies a more specific histology

So, we know we need to have both the histology on the pathology report as well as the immunophenotyping and/or genetic testing stated to be positive to assign code 3 for Diagnostic Confirmation. The next challenge is interpreting immunophenotyping test results to figure out whether or not we can use those results to code Diagnostic Confirmation! I'll review a couple of examples to demonstrate the appropriate use of codes 3 vs 1.

- In SINQ 20170006 (Figure 1), the lymph node excision diagnosis is diffuse large B-cell lymphoma (DLBCL) and flow cytometry was positive for CD antigens (immunophenotyping) 20, 22, and 30. According to the Heme Manual, under the Immunophenotyping section for DLBCL (9680/3) several other CD antigens often test positive in DLBCL but in this case they did not. However, the fact that the flow cytometry studies were positive for several antigens common to DLBCL is good enough to indicate the DLBCL was confirmed both histologically by immunophenotyping. Diagnostic confirmation should be coded to 3 for this case.

Figure 1
Diagnostic Confirmation
Code 3 Example
Question: 20170006

Question:

Heme & Lymphoid Neoplasms/Diagnostic confirmation--Lymphoma: To code 3 in Diagnostic Confirmation, does the genetic testing need to confirm a specific histology or is it enough that it simply rules out others?

Discussion:

For example, pathology states: Right axillary lymph node, excision: Diffuse large B-cell lymphoma (DLBCL) (see note). COMMENT: FISH studies were performed that were negative for BCL-6, c- Myc/IgH, CCND1/IgH and IgH/BCI-2 gene rearrangement, ruling out the most common forms of double-hit lymphoma. Flow cytometry studies demonstrated positivity for CD45, CD20, HLA-Dr, CD19, CD11c, CD22, CD30, CD38, CD79b, and FMC7. Low positivity was seen for CD5. No reactivity was seen for CD10, CD23, CD25, CD103 or CD123.

Answer:

Both histologic plus immunophenotyping or genetic testing should be positive to assign code 3 for Diagnostic Confirmation. The Hematopoietic and Lymphoid Neoplasm Coding Manual Diagnostic Confirmation instructions state, assign 3 for cases positive for neoplasm being abstracted (including acceptable ambiguous terminology and provisional diagnosis) AND Immunophenotyping, genetic testing, or JAK2 is listed in the Definitive Diagnosis in the Heme DB AND a) Confirms the neoplasm OR b) Identifies a more specific histology (not preceded by ambiguous terminology). Because the patient was diagnosed with DLBCL by histology, and flow cytometry was positive for CD antigens (immunophenotyping) 20, 22, and 30 for DLBCL, code 3 is appropriate.

- Per the Definitive Diagnostic Methods section in the Heme Manual, mantle cell lymphoma is confirmed using genetic testing, histologic confirmation, and immunophenotyping. The final diagnosis on a pathology report for a cecum resection is mantle cell lymphoma. Per the comment on the report, "Although CD5 is negative, and there is patchy weak staining for CD10, the findings support a diagnosis of mantle cell lymphoma."

Figure 2
Heme Database
Mantle Cell Lymphoma

Definitive Diagnostic Methods

Genetic testing
Histologic confirmation
Immunophenotyping

Immunophenotyping

BCL2 positive
BCL6 negative
CD5 positive
CD10 negative
CD23 negative or weakly positive
CD43 positive
Cyclin D1 expression
FMC7 positive
IgM/IgD expression
IRF4 positive
MUM1 positive

It's true the resection confirmed mantle cell lymphoma histologically. However, the immunophenotyping studies showed CD5 negativity and "patchy weak staining for CD10." Under the Immunophenotyping section, mantle cell lymphoma is CD5 positive and CD10 negative. See Figure 2. There is no clear statement in the study that the CD10 stain was negative. The expression "patchy weak staining" doesn't cut it because it may imply a weakly positive result (e.g., CD10+). There is a guideline for handling "patchy weak staining" terminology in the Heme Manual (check the Diagnostic Confirmation Coding Instructions, Code 3, Rule 1b, Note 2) that states we are not to use code 3 for positive immunophenotyping when the result is preceded by the expression "patchy weak staining."

Therefore, in this case, the immunophenotyping studies did not confirm mantle cell lymphoma because CD5 negativity is not listed under the

Immunophenotyping section in the Heme DB for this neoplasm; the CD10 result cannot be considered because of the way it was described. This case of mantle cell lymphoma should have Diagnostic confirmation coded 1 (positive histology).

Note that the code for Diagnostic Confirmation can be changed at any time. So, if a neoplasm originally confirmed by histology only (Code 1), later has immunophenotyping or genetic testing that confirms a more specific neoplasm **and the Heme Manual M Rules confirm this is the same primary**, the histology can be coded to the more specific histology and the Diagnostic Confirmation should be changed to Code 3.

Using Diagnostic Confirmation Code 5

Diagnostic confirmation Code 5 (Positive laboratory test/marker study) is used only when the diagnosis of cancer is based on laboratory tests, tumor marker studies, genetics or immunophenotyping that are clinically diagnostic for that specific cancer. The diagnostic laboratory tests that might apply for a particular histology are listed in the Hematopoietic DB Definitive Diagnostic Methods section for that disease process. Be aware that this Diagnostic Confirmation code is **rarely** used because if there was no provisional diagnosis or clinical suspicion of cancer from a histologic specimen, immunophenotyping or genetic testing would likely not be done. These tests are typically performed to identify a more specific histologic type after either a cytologic or histologic specimen is positive or suspicious for a reportable heme/lymphoid neoplasm.

However, in some cases the Diagnostic Confirmation Code 5 does apply. For example, when a CT scan is consistent with multiple myeloma (9732/3) and 24-hour urine protein is elevated with the presence of Bence-Jones kappa, Code 5 is used because the diagnosis is based on the positive Bence-Jones lab test, which is listed as one of the definitive diagnostic methods in the Heme DB for multiple myeloma. Code 3 does not apply because there is no histologic confirmation of the myeloma.

Using Diagnostic Confirmation Code 8

Diagnostic confirmation Code 8 (Clinical diagnosis only; other than lab test, marker study, direct visualization or imaging) is assigned when the diagnosis is based on the healthcare professional's expertise, combined with the information from equivocal or negative tests and the clinical presentation. This code is often used when the histology is not stated definitely using any microscopic method nor is the histology stated definitely in a lab test, marker study, scan or because it was directly visualized.

Code 8 is appropriately used for cases when a reportable diagnosis reached through a process of elimination known as a **diagnosis of exclusion**. This term refers to a diagnosis of a reportable disease process reached when its presence cannot be established with complete confidence from the patient's history or through examination or testing. For instance, Diagnostic Confirmation Code 8 applies to a case in which a bone marrow biopsy shows anemia NOS and physician notes state that the patient's overall clinical appearance of hypercalcemia, fever, and anemia is consistent with Myelodysplastic Syndrome, NOS (9989/3).

One of the more common Diagnostic Confirmation coding mistakes involves incorrectly coding some of the JAK-2 positive neoplasms. Many of us use code 5 (Positive laboratory test/marker study) inappropriately when we have only a clinical assessment that a patient has a reportable disease process PLUS a test result indicating the disease process is JAK-2 positive. According to

the Heme Manual, a JAK-2 positive result alone is never considered diagnostic of a specific hematologic neoplasm. In a situation where you have only a clinical diagnosis of a myeloproliferative neoplasm (MPN) such as polycythemia vera (PV), essential thrombocythemia (ET), or agnogenic myeloid metaplasia (primary myelofibrosis, PMF) and a JAK-2 positive test result, code Diagnostic Confirmation to 8. See Figure 3 to read SEER's SINQ 20180010 answer that supports this coding decision.

Importance of correctly coding Diagnostic Confirmation

It is important to correctly code the Diagnostic Confirmation field because it represents the method by which provisional and final diagnoses are made. Understanding how to interpret genetic and immunophenotyping studies are critical to the classification of a specific hematopoietic neoplasm and their treatment; therefore, it is necessary to document these with an accurate code. While working on hematopoietic and lymphoid cases, we need to be extra cautious and make sure we consider all the test reports and pathology reports as well as the terms used by the physician to describe the neoplasm.

Figure 3
Diagnostic Confirmation
Code 8 - Yes/Code 5 - No
Question: 20180010

Question:

Diagnostic confirmation—Heme & Lymphoid Neoplasms: Is Diagnostic Confirmation coded as 5 (positive laboratory test/ marker study) or code 8 (clinical diagnosis only) for a case that has a positive JAK2 mutation, and based on the results of the JAK2, the physician diagnosed the patient with polycythemia vera? There were no blood smears or bone marrow biopsies done.

Answer:

Assign diagnostic confirmation code 8 for a clinical diagnosis only. Code 5 is not correct in this case because JAK2 is not definitive for any specific hematopoietic neoplasm. The physician uses JAK2 info combined with all of the other facts for the case to make the diagnosis.