

Implementing ICD-O-3: Impact of the New Edition

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Abstract The *International Classification of Diseases for Oncology, third edition (ICD-O-3)* will be implemented in North America with cases diagnosed January 1, 2001, and after. The introduction of ICD-O-3 provides an opportunity to remind coders, registrars, and others of the intricacies of neoplasm coding. This technical overview of the new edition describes the process of revising a standard reference, lists and discusses terms that have changed behavior code (making them reportable diagnoses), illustrates features and enhancements in the new edition, and revisits important points about neoplasm coding that have not changed in the process of developing a new reference manual.

Background

For more than twenty-five years, the *International Classification of Diseases for Oncology (ICD-O)* has been the standard by which neoplasms have been coded throughout the world. The ten digit code describes where the tumor arose (a four character topography code for the primary site), what the tumor is (a four digit histology code for the cell type), how it behaves (a one digit code for malignant, benign, and so forth) and how aggressive it is (a one digit code for differentiation or grade). The first edition of ICD-O was published in 1976, and a substantial revision – primarily of the topography codes – was published in 1990.

Immense changes have occurred over the past decade in techniques for diagnosing neoplasms. As a result, pathologists have been able to provide much more specific information about certain cancers. In particular, cytogenetic techniques have added considerably to the body of knowledge about malignant lymphomas and leukemias. In some cases, the names of the diseases have changed to reflect the additional information. As a consequence, cancer registries and pathology departments using the *International Classification of Diseases for Oncology, second edition (ICD-O-2)* [1] have been unable to code these new entities satisfactorily.

Responding to requests for assignment of new code numbers for these entities, in 1998, the International Agency for Research on Cancer (IARC), the cancer division of the World Health Organization, gathered a task force to assess whether a revision or new edition of ICD-O was necessary. It was initially thought that only the lymphomas and leukemias would be revised. However, when questionnaires sent to every national registry in the world indicated that new diagnostic terms had been identified in all categories of neoplasms, it was decided to update the entire book. It was not necessary to revise the topography section of ICD-O-2, since it is based on the *International Classification of Diseases, tenth revision* [2].

In addition, there was a desire that the next edition of ICD-O be compatible with other World Health Organization publications such as the series *Histological Typing of Tumours*, known to pathologists around the world as the Blue Books [3]. In a coordinated effort with the editors of the Blue Books, all terms in existing fascicles were reviewed to assure that the histologic terminology was included in ICD-O-3. Further, fascicles in preparation have been reviewed to assign ICD-O-3 codes to the terms listed in their type lists.

ICD-O-3 underwent a limited but comprehensive field trial last summer. The editors met in October 1999 and again in January 2000 to finalize the lists of terms and codes and to add any additional terms found

during the field trial. Additional committee work has been conducted by conference call and electronic mail.

ICD-O-3 will be implemented in North America effective with cases diagnosed on or after January 1, 2001. The advent of a new edition of ICD-O offers the opportunity to review important aspects of coding guidelines that have not changed, to highlight the enhancements provided by the editors, and to underscore what is new and what is different in the third edition.

WHAT' S NEW

Terms changing to malignant behavior

Perhaps the most important change between ICD-O-2 and ICD-O-3 involves a small number of disease entities that have changed behavior codes. Eleven diagnoses are changing from borderline to malignant to reflect current medical understanding about the behavior of these neoplasms (Table 1). They include all of the refractory anemia types, polycythemia vera, a number of other hematopoietic diseases, and papillary meningioma,. The change from behavior code 1 (borderline) to behavior code 3 (malignant) will make these diseases reportable to cancer registries that do not currently require their collection. The impact on cancer registries that must add these cases to their databases is not believed to be substantial. SEER estimates 5,000 to 10,000 new cases per year in the U.S., about the frequency of pharynx, gallbladder, or testicular cancer or Hodgkin's disease or chronic lymphocytic leukemia. It will, however, be necessary to add new codes to current casefinding code lists in order to identify these newly reportable diagnoses from the medical record disease indices.

Terms changing to borderline behavior

Five neoplasms previously coded as malignant (/3) will revert to borderline status: the cystadenomas of the ovary (Table 2). A careful review of survival rates for the cystadenomas, which had been considered malignant while ICD-O-2 was in effect, indicated that their behavior was much closer to benign rather than malignant and the overall survival rate for them was nearly 100%. In most registries, these neoplasms will now be considered non-reportable and it will not be necessary to collect them. Cystadenomas of the ovary collected during the period when ICD-O-2 was in effect may be deleted from registry databases at the discretion of the database manager or standard-setting agency. One additional term, pilocytic or juvenile astrocytoma, and one obsolete term (spongioblastoma, NOS) will also revert to borderline behavior. However, for the sake of continuity, pilocytic astrocytoma will continue to be collected in many registries for the next few years.

Table 3 lists other tumors that have changed to or from borderline status; these diagnoses are considered non-reportable by most cancer registries.

New codes, terms and synonyms

About 780 morphology terms have been added to ICD-O-3. Over 500 terms were added as new terms or synonyms to existing codes in the 8000-9580 range, and over 200 terms and synonyms were added to the leukemia and lymphoma section. In the process of reorganizing the lymphomas and leukemias, a number of terms were moved to different codes or combined with other codes, but numbers were never re-used to avoid the potential confusion of multiple meanings over time for a single code. New code numbers were added representing 220 new morphologic entities and 400 terms and synonyms. Approximately 380 new synonyms have been added to existing codes. Complete lists of morphology code changes, behavior code changes, new terms and synonyms for existing codes are available from the SEER Program website, www.seer.cancer.gov/administration.

Although the purpose of the new edition was to include terms from new classification systems, it was also necessary that older terminology be retained for reference by pathology departments and cancer registries where data has been collected over several decades and changes in coding manuals. As a result, it has not always been possible to assign codes to new entities in the same sequence in which they appear in their original classification. For example, the term »follicular lymphoma, grade 2« is actually a new synonym for the existing code 9691/3, »malignant lymphoma, follicle center cell, mixed small cleaved and large cell.« Grade 1 follicular lymphoma is a new synonym for 9695/3, »malignant lymphoma, follicle center cell, small cleaved.« It appears that the terms with grade descriptors are out of sequence in the numerical list, but in fact, these terms are now the preferred names for previously identified lymphomas. Additionally, in certain sections of the numerical list, there was no room to add new codes in sequence and codes had to be inserted where numbers were available. Thus despite its name, the *International Classification*, the third edition of ICD-O should be considered a coded nomenclature rather than a true classification of disease entities.

In many instances, there is more specific designation of the preferred behavior of an NOS term (Not Otherwise Specified or nonspecific) when the word benign, borderline, or malignant is not part of the diagnostic phrase. For example, if the diagnostic statement says simply »teratoma,« this is to be coded as 9080 with a behavior code of /1. If the diagnostic statement were to say »malignant teratoma,« the code would be 9080/3.

To reflect contemporary pathology practice and terminology, a number of revisions and enhancements have been made to ICD-O-3. More acronyms have been included as synonyms in the numeric list and in the alphabetic index, such as DCIS (ductal carcinoma in situ) and PNET (primitive neuroectodermal tumor). Table 4 lists some of the more lengthy or obscure acronyms and their complete definitions.

In addition to the very specific codes for newly-identified disease entities, several helpful codes have been added for non-specific diagnoses, such as 8046/3, non-small cell carcinoma and 9861/3, acute non-lymphocytic leukemia (ANLL). Although these are not »good« diagnoses according to the pathologists, registrars see them frequently on cytology reports and death certificates, so these terms were included in ICD-O-3. Also, having a code for these disease entities allows registrars to »mark« these cases for case follow-back for a more specific diagnosis. For example, following back on a non-small cell carcinoma case coded to 8046/3 may afford the opportunity to code to a more specific non-small cell carcinoma such as adenocarcinoma or large cell carcinoma.

Some terms listed in ICD-O-3 have been marked as obsolete [obs]. This descriptor is intended to discourage the use of such a term for a new diagnosis when better diagnostic terms are available, yet to serve as a reference when such a diagnosis is noted during research using historical data. Some terms are older names for neoplasms that have been more specifically described, for example hepatoma [obs] which is now described as hepatocellular carcinoma with new codes for four subtypes. Others are truly archaic, such as lymphosarcoma (first described in the 1890s, although the term is still used in veterinary medicine). In many cases, obsolete terms which had specific codes in ICD-O-2 have been moved to the »Not Otherwise Specified« category for the disease.

Eponymic terms (diseases named after a person) will be listed in ICD-O-3 in their European format, without the apostrophe s (’s). Both the numeric and alphabetic lists will show Klat skin tumor and Hodgkin lymphoma, for example.

Introduction and Coding Guidelines Enhanced

In response to comments from beginning coders and non-English speaking users of ICD-O, the coding instructions in the front of the book have been rewritten, clarified, and made more user-friendly. Page

numbering throughout the book is sequential, avoiding the lower case roman numerals in the introductory section that have mystified so many people learning to use the manual. Many examples extracted from the text of ICD-O-2 have been placed in tables to make them clearer.

Description of the structure of the codes has been separated from the coding rules. The coding rules have been reorganized (first topography rules and then morphology rules) and identified differently (by letter rather than number) to avoid confusion with rules in previous editions. The concepts expressed in the rules themselves have not changed, but the wording has been simplified to meet the needs of beginning coders and in areas where English is not the primary language. Sentence structure has been revised to use active verbs and to simplify complex sentences as the example in Table 5 shows. All coders are encouraged to re-read the Introduction and Coding Guidelines to familiarize themselves once again with the basic concepts of ICD-O coding.

The alphabetic index has been enhanced by adding subheaders to long lists of terms, such as those listed under carcinoma and tumor, to make looking up codes faster and easier (Table 6). This enhancement is an expansion of the format used for lymphomas and leukemias in the previous edition.

Lymphomas and Leukemias

The field of lymphoma and leukemia research is well known for the variety of classification systems that exist for these diseases. In the second edition of ICD-O, the lymphoma section was largely based on the Working Formulation [4], which was a means of translating terminology among the Rappaport, Kiel, and Lukes-Collins classifications. The terminology from all these classifications was included in ICD-O-2. In 1994, a new classification of lymphoid neoplasms, called the Revised European-American Lymphoma (REAL) classification [5], was published. Rather than group lymphomas by the physical characteristics of their cells (such as follicular or diffuse morphology), the REAL classification grouped diseases of the blood and lymphatic tissues along their cell lines. This was a major shift in hematologic thinking, and resulted in a clinical advisory committee meeting in 1997 to reach consensus on the usefulness of the new classification system. The result was the World Health Organization Classification of Neoplastic Diseases of the Hematopoietic and Lymphoid Tissues [6], published in 1999, which is now accepted as the international standard for describing this vast array of diseases. To accommodate the new terminology, the lymphoma and leukemia sections of ICD-O-3 were similarly revised and grouped along cell lines. Terms from the previous classifications were sorted and grouped by their cell derivation, and new terms were added to the lists. Table 7 shows how the three-digit ICD-O-3 morphology groupings reflect the organization of the WHO Classification.

This division of diseases along cell lines led to some additional coding issues, because the consensus at the 1997 WHO conference was that some lymphomas and leukemias are the same disease with different presentations. For example, the WHO Classification lists B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma as a single entity, the same disease at different stages. The hematopathologists on the ICD-O-3 development committee recommended a single code number to represent the disease. However, since ICD-O is a subset of ICD-10 and ICD-10 is used to code mortality throughout the world, if a single ICD-O-3 code were used, there would be no way to determine whether a death was due to lymphoma or leukemia which are coded separately in ICD-10. As a result, it was necessary to retain separate codes for chronic lymphocytic leukemia and small lymphocytic lymphoma and link them. Thus, for the first time in ICD-O editions, single disease entities are listed in two different categories and cross-referenced. Table 8 lists other pairs of disease entities that are considered different in ICD-10 and the same in the WHO classification. The topographic or primary site code for pairs such as these depends on where the disease is diagnosed: if disease is diagnosed only in the blood or bone marrow, code the

primary site to C42.1, bone marrow. If the diagnosis is made on any other tissue, code to the tissue involved, usually the lymph node.

One of the decisions reached at the 1997 WHO conference was to recognize the importance of defining and naming leukemias by the chromosomal changes in cancerous cells. As a result, the third edition of ICD-O includes cytogenetic qualifiers for certain hematologic terms (Table 9) that may be confusing to coders and abstractors. For example, the preferred term for 9866/3, acute promyelocytic leukemia, includes the suffix t(15;17)(q22;q11-12). Normal human cells have 23 pairs of autosomal chromosomes, numbered 1 to 23, and two sex chromosomes, labeled AX@ or AY.@ In cytogenetic shorthand, *t* means translocation or a reciprocal exchange of genetic material between two chromosomes. So in this example, t(15;17)(q22;q11-12) means that in cancerous cells, the material from chromosome 15 on its long arm (*q* means long arm) in region 22 is swapped with the material on chromosome 17 on its long arm in the region between 11 and 12.

Acute promyelocytic leukemia is also described by its molecular abnormality, PML/RAR-alpha. In the WHO Classification, the entity is listed as acute promyelocytic leukemia (AML with t(15,17)(q22;q11-12), PML/RAR-alpha. Since this complete description may not be included in the final diagnosis on a pathology report, the editors of ICD-O-3 thought it best to break such terms into their cytogenetic and molecular elements, as the example in Table 9 shows.

The French-American-British (FAB) classification of leukemias, which was not in ICD-O-2 but was commonly used by pathologists, has been folded into the WHO Classification of hematopoietic diseases. The M0 through M7 terminology of the FAB classification has been included in ICD-O-3 for reference, since these terms are still in widespread use.

WHAT'S NOT NEW (BUT IMPORTANT NONETHELESS)

The Matrix Principle

Although many more specific diagnoses and code-behavior combinations have been added to ICD-O-3 to accommodate electronic reference files, the matrix principle that was introduced in the first edition of ICD-O should be reinforced as another coding tool. The matrix concept is the rule that says it is permitted to change the behavior code of a reported diagnosis so that it truly reflects what the pathologist describes as the behavior. Thus if a pathologist reports a diagnosis of "malignant adrenal rest tumor," the coder may take the 4-digit histology code for adrenal rest tumor, 8671, and change the behavior code /0 to /3. While this will probably generate an edit check or advisory message in a computerized registry database, the system should have the ability to over-ride the message and save the case with a histology-behavior combination that is not printed in ICD-O-3.

Old Terms, New Meanings

Because of changes in the terminology used by pathologists, there is some potential for confusion by non-physicians and coders. For example, pathologists use the term "grade" as a synonym for "type" or "category." Registrars, on the other hand, recognize the term "grade" as an indicator of cell differentiation which is coded in the 6th digit of the ICD-O morphology code. Therefore, it is important to recognize when the term "grade" refers to category and when it refers to biologic activity.

Similarly, to a pathologist, the terms "adult" and "mature" describe cell characteristics rather than the age of the patient. "Transitional" has two meanings--it could be a cell type, or it could be a neoplasm that is converting to something else. Careful communication and explanation of new terms is necessary to assure that the new information is coded correctly.

Revisions and Exceptions to Common Guidelines

Advances in diagnostic technology and the classification of histologic subtypes of certain diseases have also affected ICD-O-3. For example, many different subtypes have been described for duct cell carcinoma, and several subtypes of kidney cancer have also been reported in the Mainz classification of renal tumors. Some of the subtypes have been assigned lower code numbers than the NOS term and this may affect ICD-O's "code to the highest code as it is generally more specific" rule. Code the subtype of the tumor, even if it is a lower number than the NOS term. For example, "ductal carcinoma, cribriform type" is a new synonym for 8201/3, cribriform carcinoma.

For breast cancer cases with multiple subtypes, two new codes have been included in ICD-O-3. Use code 8523/_ (in situ or invasive as appropriate) when there is a diagnosis of duct carcinoma mixed with more than one subtype, such as ductal carcinoma with elements of cribriform, mucinous and lobular carcinoma. Code 8524/_ when there is a diagnosis of lobular carcinoma mixed with more than one other type of carcinoma.

In 1998, the World Health Organization and the International Society of Urologic Pathologists published a consensus paper about describing the preferred terminology for various types of bladder tumors and these terms have been added to ICD-O-3. There are also many new synonyms for papillary bladder tumors. It is important to note that the code for non-invasive papillary transitional cell carcinoma, 8130/2, will be printed in ICD-O-3 and will not have to be created from the matrix principle. This should relieve some of the edit messages registrars receive when coding a non-invasive bladder tumor.

In addition, the terminology differences between benign non-reportable and malignant reportable diagnoses are graying. The registry rule of thumb that states "if the term contains a cancer word like 'leukemia' or 'sarcoma' or 'astrocytoma,' the case is malignant and reportable" no longer applies 100% of the time. As pathologists have identified very indolent varieties of certain tumors, they have assigned them behavior codes of /1, uncertain whether benign or malignant. Conversely, there are a number of terms that do not contain a common cancer word that are indeed malignant. Table 10 shows examples of these potentially confusing terms of malignant and borderline behavior.

Conversions

As mentioned previously, a list of all changes between ICD-O-2 and ICD-O-3 is available for downloading from the "Administration" section of the SEER web site, www.seer.cancer.gov. Cancer registry software programmers should download this file to begin updating the reference files in their application programs.

The Uniform Data Standards Committee of the North American Association of Central Cancer Registries has approved two new data fields to store new ICD-O-3 data or codes converted from ICD-O-2: "histologic type ICD-O-3" and "ICD-O-3 conversion code." Histology codes prior to 1/1/2001 will be maintained in the original field. In addition, these codes should be converted into the new "histologic type ICD-O-3" field and the flag set in the "ICD-O-3 conversion code" field to indicate that the data were converted. In this way, historical data can be preserved while analysis of data over time can be performed on a single data element. In addition, the editors of ICD-O-3 are preparing standardized conversion programs and reference tables of ICD-O-2 to ICD-O-3 and ICD-O-3 to ICD-O-2 that software developers may incorporate into their conversion routines. These will be available from the SEER Program; announcements of their availability will be broadcast widely.

During the conversion from ICD-O-2 to ICD-O-3, most terms will copy through without review or change. A few terms will change number during the conversion, and a few terms that have been split

apart will require manual review of supporting text fields or original documents. In general, these are very rare diagnoses about which more has been learned, and many of the terms are benign diseases that are not included in most registries. For example, thymoma, NOS will be separated from benign thymoma and will become a borderline term. Thymic carcinoma moves to a new number discrete from malignant thymoma; there were fewer than 300 cases in the SEER database diagnosed between 1992 and 1997. Recoding the parosteal and juxtacortical osteosarcomas will require review of about 50 cases in SEER areas.

Other diagnoses can be recoded without manual intervention simply by reviewing the primary site associated with the diagnosis. For example, stromal sarcomas will be separate from endometrial stromal sarcomas and can be identified by any primary site code that is not C54.1, endometrium. Extramedullary plasmacytomas are non-marrow primary sites, whereas the plasmacytomas from which they are being separated are always coded to bone marrow (C42.1).

At least one diagnosis should be reviewed for quality control purposes as a result of the introduction of the WHO Classification of Hematopoietic Diseases. In addition to the four Hodgkin's disease (now called Hodgkin lymphoma)--cell types in the Rye Classification (nodular sclerosis, mixed, lymphocyte predominant and lymphocyte deplete), there is a newer disease entity: nodular lymphocyte predominance Hodgkin lymphoma. This new entity is different from what is now called "classic" lymphocyte predominance Hodgkin's disease, which has been re-named lymphocyte-rich. Since no code for nodular lymphocyte predominance Hodgkin lymphoma existed in ICD-O-2 and the diagnosis has been recognized for nearly two decades, it would be advisable to review morphology text fields to assure that these different diseases are correctly coded in ICD-O-3.

Training Opportunities

The SEER Program is developing a self-instructional web-based module on the changes that will occur with the implementation of ICD-O-3. It is expected that continuing education hours will be granted for this module (planned for release in the fall). In addition, a satellite broadcast similar to the surgery codes telecast is scheduled for January after registrars have had the opportunity to obtain copies of the new manual. Announcements of the web training, satellite broadcast, and availability of the books for purchase will be widely disseminated through media accessed by registry professionals, including the web sites for SEER (www.seer.cancer.gov), NCRA (www.ncra-usa.org) and NAACCR (www.naacr.org), newsletters and journals such as this.

Obtaining the New Manual

WHO has set the price for single copies of ICD-O-3 at \$54.00. There is a 30% discount (\$37.80) for 30 to 999 copies shipped to a single address, and a 60% discount (\$21.60) for 1000 or more copies shipped to a single address. As of this writing, a specific publication date has not been established. The ICD-O-3 Editorial Committee has requested that WHO publish the book in hardbound, softbound and electronic versions. These decisions will be made by the publisher. ICD-O-3 will be available from the World Health Organization's North American distributor, WHO Publications Center USA, 49 Sheridan Avenue, Albany, NY 12210, and other sources.

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Table 1. Terms that Changed to Malignant Behavior

ICD-O-2		ICD-O-3
Code	Primary term as it appeared in ICD-O-2	Code
8931/1	Endolymphatic stromal myosis (C54.1)	8931/3
9134/1	Intravascular bronchial alveolar tumor (C34._)	9133/3
9538/1	Papillary meningioma	9538/3
9950/1	Polycythemia vera	9950/3
9960/1	Chronic myeloproliferative disease, NOS	9960/3
9961/1	Myelosclerosis with myeloid metaplasia	9961/3
9962/1	Idiopathic thrombocythemia	9962/3
9980/1	Refractory anemia, NOS	9980/3
9982/1	Refractory anemia with sideroblasts	9982/3
9983/1	Refractory anemia with excess blasts	9983/3
9984/1	Refractory anemia with excess blasts in transformation	9984/3

Table 2. Terms that Changed from Malignant to Borderline

ICD-O-2 Code	Primary term as it appears in ICD-O-3	ICD-O-3 Code
8442/3	Serous cystadenoma, borderline malignancy (C56.9)	8442/1
8451/3	Papillary cystadenoma, borderline malignancy (C56.9)	8451/1
8462/3	Serous papillary cystic tumor of borderline malignancy (C56.9)	8462/1
8472/3	Mucinous cystic tumor of borderline malignancy (C56.9)	8472/1
8473/3	Papillary mucinous cystadenoma, borderline malignancy (C56.9)	8473/1
9421/3	Pilocytic astrocytoma (C71._)	9421/1
9422/3	Spongioblastoma, NOS (C71._)	9421/1

Table 3. Terms that Changed to or From Borderline Behavior

ICD-O-2 Code	Primary term as it appears in ICD-O-3	ICD-O-3 Code
8152/0	Glucagonoma, NOS (C25._)	8152/1
8580/0	Thymoma, NOS (C37.9)	8580/1
8640/0	Sertoli cell tumor, NOS	8640/1
9506/0	Neurocytoma	9506/1
8261/1	Villous adenoma, NOS	8261/0
8361/1	Juxtaglomerular tumor (C64.9)	8361/0
8823/1	Desmoplastic fibroma	8823/0
9080/1	Mature teratoma	9080/0

Table 4. Examples of Acronyms included in ICDO-3

Acronym	Complete term incorporating acronym
SETTLE	Spindle Epithelial Tumor with Thymus-Like Element
CASTLE	CArcinoma Showing Thymus-Like Element
MPNST	Malignant Peripheral Nerve Sheath Tumor
MALT, SALT, BALT	-Associated Lymphoid Tissue (Mucosa, Skin, Bronchial)
PIN, AIN, DIN	Intraepithelial Neoplasia (Prostate, Anal, Ductal)
PNET, CPNET, PPNET	NeuroEctodermal Tumor (Primitive, Central Primitive, Peripheral Primitive)
RA, RARS, RAEB, RAEB-T	Refractory Anemia (with Ringed Sideroblasts, with Excess Blasts, with Excess Blasts in Transformation)

Table 5. Example of Revised Wording of Coding Rules

OLD WORDING (Rule 6)

If a diagnosis indicates two different degrees of grade or differentiation (e.g., "well and poorly differentiated" or "grades II-III"), code to the higher grade.

NEW WORDING (Rule G)

Grading or differentiation code: Assign the highest grade or differentiation code described in the diagnostic statement.

Table 6. Example of Enhanced Index Entry

<i>Adenoma, continued</i>	
M-8640/1	testicular
M-8190/0	trabecular
M-8336/0	trabecular, hyalinizing (C73.9)
	Tubular
M-8211/0	NOS
M-8210/3	adenocarcinoma in
M-8210/2	adenocarcinoma in situ in
M-8640/1	Pick
	Tubulovillous
M-8263/0	NOS
M-8263/3	adenocarcinoma in
M-8263/2	adenocarcinoma in situ in
M-8263/0	villoglandular
	Villous
M-8261/0	NOS
M-8261/3	adenocarcinoma in
M-8261/2	adenocarcinoma in situ in
M-8322/0	water-clear cell (C75.0)
M-9110/0	Wolffian duct

Table 7. Lymphoma and Leukemia Groupings

959	Lymphoma, NOS
965-966	Hodgkin lymphoma
967-969	Mature B-cell lymphomas
970-971	Mature T- and NK-cell lymphoma
972	Precursor cell lymphoblastic lymphoma
973	Plasma cell tumors
974	Mast cell tumors
975	Histiocyte and accessory lymphoid cell tumors
976	Immunoproliferative diseases
980	Leukemia, NOS
982-983	Lymphoid leukemia
984-993	Myeloid leukemia
994	Other leukemias
995-996	Chronic myeloproliferative disorders
997	Other hematologic disorders
998	Myelodysplastic syndromes

Table 8. Cross-referenced Hematopoietic Disease Entities

- 9670/3 Malignant lymphoma, small B lymphocytic, NOS (*see also M-9823/3*)
9823/3 B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma (*see also M-9670/3*)
- 9671/3 Malignant lymphoma, lymphoplasmacytic (*see also M-9761/3*)
9761/3 Waldenstrom macroglobulinemia (C42.0) (*see also M-9671/3*)
- 9675/3 Malignant lymphoma, mixed small and large cell, diffuse [obs] (*see also M-9690/3*)
9690/3 Follicular lymphoma, NOS (*see also M-9675/3*)
- 9687/3 Burkitt lymphoma, NOS (*see also M-9826/3*)
9826/3 Burkitt cell leukemia (*see also M-9687/3*)
- 9727/3 Precursor cell lymphoblastic lymphoma, NOS (*see also M-9835/3*)
9835/3 Precursor cell lymphoblastic leukemia, NOS (*see also M-9727/3*)
- 9728/3 Precursor B-cell lymphoblastic lymphoma (*see also M-9836/3*)
9836/3 Precursor B-cell lymphoblastic leukemia (*see also M-9728/3*)
- 9729/3 Precursor T-cell lymphoblastic lymphoma (*see also M-9837/3*)
9837/3 Precursor T-cell lymphoblastic leukemia (*see also M-9729/3*)

Table 9. Examples of Cytogenetic Descriptors

9866/3	Acute promyelocytic leukemia t(15;17)(q22;q11-12) Acute promyelocytic leukemia, PML/RAR-alpha Acute promyelocytic leukemia, NOS FAB M3
9875/3	Chronic myelogenous leukemia, BCR/ABL positive Chronic granulocytic leukemia, t(9;22)(q34;q11) Chronic granulocytic leukemia, Philadelphia chromosome (Ph1) positive

Table 10. Examples of Terms with Confusing Behavior Indicators

Borderline Terms that Don't Sound Borderline (Not Reportable)

Metastasizing leiomyoma

T-cell and NK-cell large granular lymphocytic leukemia

Superficial well-differentiated liposarcoma

Well-differentiated liposarcoma of superficial soft tissue

Juvenile melanoma (benign)

Malignant Terms that Don't Sound Malignant (Reportable)

Atypical carcinoid

Rhabdoid tumor, NOS

Intratubular germ cell neoplasia

Epithelioid trophoblastic tumor

Mixed pineal tumor, transitional pineal tumor

Atypical teratoid/rhabdoid tumor

Pagetoid reticulosis

Generalized Langerhans cell histiocytosis

Agnogenic myeloid metaplasia

Heavy chain disease

Immunoproliferative small intestinal disease