ICD-O-3 Errata and Clarifications

May 22, 2001

Errata and Clarifications for two versions of the *International Classification of Diseases for Oncology, third edition (ICD-O-3)* are included in this document. The hardcover version was sent to the World Health Organization printer in June 2000. After the masters for the hardcover version were printed, a number of discrepancies between the morphology numeric list and the index were identified and corrected. The softcover version, subtitled "U.S. Interim Version," which was produced by the U.S. government for use in the United States and sent to the printer in November 2000, included these corrections. Subsequent to the delivery of both versions of ICD-O-3, a number of additional errata were identified. ALL errata are included in this document.

Please note that the majority of the errata are in the index. If there is a discrepancy between the numeric list and the index, the numeric list is considered the authority. Only four codes have been identified as incorrect in the index; only one code was incorrectly listed as a borderline (/1) tumor when it should have been malignant (/3). An additional term was incorrectly listed as malignant when the correct code is borderline. However, because each term is indexed under each of the words in the phrase, the incorrect codes must be corrected in several places in the index. The codes that MUST be corrected because they affect reportability are listed in Table 1. Editorial changes to make the index consistent with the numeric list, such as adding or deleting a suggested site code or to correct the alphabetic sequence, are listed in Table 2.

The hardcover version of ICD-O-3 is available for purchase from World Health Organization publication centers. An electronic version of ICD-O-3 will be available from WHO on CD-ROM in the summer of 2001. The WHO North American publication center is

WHO Publications Center USA
47 Sheridan Avenue
Albany, NY 12210
phone: 518-436-9686 fax: 518-436-7433 e-mail: QCORP@compuserve.com

The softcover version of ICD-O-3 is available ONLY from the SEER Program of the National Cancer Institute (see address below) or the National Program of Cancer Registries of the Centers for Disease Control and Prevention. DO NOT contact WHO or its publication centers to request the softcover version.

If there are any questions about this document or if additional discrepancies are identified, please notify

April Fritz, ICD-O-3 Support
SEER Program, National Cancer Institute
Suite 504, MSC 8316
6116 Executive Blvd
Rockville, MD 20852
e-mail: april.fritz@nih.gov fax: 301-496-9949

The following table lists the following items to aid in locating a line to be corrected

PG (Page number), COL (Column on page), LINE (Line number–includes each vertical space counted as one line regardless of height), HC (an x means the change affects hardcover version), SC (an x means the change affects softcover version), TERM (Code number and term), ACTION (description of how to change the entry).

Please make the following corrections in your copy of the *International Classification of Diseases for Oncology, third edition*..

TABLE 1. The following corrections affect reportability of malignancies. These changes MUST be made in the Index.

PG	COL	LINE	HC	SC	TERM	ACTION
94	2	37	x	x	9421/1 Pilocytic astrocytoma (C71._) [including all synonyms]	*Add note:* In North America, report as 9421/3
113	2	27	x	x	M-9421/1 Astrocytoma, juvenile (C71._)	*Add note:* In North America, report as 9421/3
113	2	30	x	x	M-9421/1 Astrocytoma, pilocytic (C71._)	*Add note:* In North America, report as 9421/3
113	2	31	x	x	M-9421/1 Astrocytoma, piloid (C71._)	*Add note:* In North America, report as 9421/3
132	2	39	x	x	M-9980/3 Cytopenia with multilineage dysplasia, refractory	***Replace code with:* M-9985/3**
135	2	35	x	x	M-9980/3 Dysplasia, multilineage, refractory cytopenia with	***Replace code with:* M-9985/3**
155	2	19	x	x	M-9421/1 Juvenile astrocytoma (C71._)	*Add note:* In North America, report as 9421/3
172	2	53	x	x	M-8734/3 Melanoma, desmoplastic, amelanotic (C44._)	***Replace code with:* M-8745/3**
173	2	37	x		M-9538/1 Meningioma, rhabdoid	***Replace code with:* M-9538/3**
177	2	27	x	x	M-9980/3 Multilineage dysplasia, refractory cytopenia with	***Replace code with:* M-9985/3**
178	1	50	x	x	M-9989/1 Myelodysplastic syndrome, NOS	***Replace code with:* M-9989/3**
189	2	46	x	x	M-9421/1 Pilocytic astrocytoma (C71._)	*Add note:* In North America, report as 9421/3
189	2	47	x	x	M-9421/1 Piloid astrocytoma (C71._)	*Add note:* In North America, report as 9421/3
192	1	28	x	x	M-9989/1 Preleukemia [obs]	***Replace code with:* M-9989/3**
192	1	29	x	x	M-9989/1 Preleukemic syndrome (C42.1) [obs]	***Replace code with:* M-9989/3**
194	1	24	x	x	M-9980/3 Refractory cytopenia with multilineage dysplasia	***Replace code with:* M-9985/3**
205	2	51	x	x	M-9989/1 Syndrome, myelodysplastic, NOS (C42.1)	***Replace code with:* M-9989/3**
205	2	56	x	x	M-9989/1 Syndrome, preleukemic (C42.1) [obs]	***Replace code with:* M-9989/3**
213	1	27	x	x	M-8473/3 Tumor, mucinous, papillary, of low malignant potential (C56.9)	***Replace code with:* M-8473/1**

TABLE 2. The following corrections are editorial and do not affect reportability. They should be made to assure uniformity between the numeric list and the index.

PG	COL	LINE	HC	SC	TERM	ACTION
7	-	2	x	x	M-9680/3 (large B-cell lymphoma)	*Replace with* M-9670/3 (diffuse small cell lymphoma)
20	-	25	x		Rule D.	*Replace with text of Rule D, page 26.*
101	1	44	x	x	9767/1 Angioimmunoblastic lymphadenopathy	*Add at end of line:* (AIL)
101	1	45	x	x	9767/1 synonym [indented]	*Add as synonym:* Immunoblastic lymphadenopathy (IBL) [obs]
110	1	54	x	x	M-9767/1 AIL (Angioimmunoblastic lymphadenopathy)	*Insert new term*
111	2	35	x	x	M-9767/1 Angioimmunoblastic lymphadenopathy	*Add at end of line:* (AIL)
121	2	39	x	x	M-8076/2 Carcinoma, epidermoid, in situ with questionable stromal invasion (C53._)	*Delete:* (C53._)
122	2	12	x	x	*In space after* M-8504/2 Carcinoma, intracystic, noninfiltrating	*Insert new term even with 'intracystic':* M-8504/3 intracystic, papillary
122	2	40	x	x	M-8072/3 Carcinoma, large cell, squamous cell, nonkeratinizing	*Add at end of line:* , NOS
123	1	34	x	x	M-8180/3 Carcinoma, mixed, hepatocellular and bile duct	*Add at end of line:* (C22.0)
123	2	35	x	x	*In space after* M-8050/2 Carcinoma, papillary, in situ	*Insert new term even with 'in situ':* M-8504/3 intracystic
124	1	49	x	x	M-8043/3 Carcinoma, small cell, fusiform cell (C34._)	*Delete:* (C34._)
124	1	50	x	x	M-8044/3 Carcinoma, small cell, intermediate cell (C34._)	*Delete:* (C34._)
124	2	18	x	x	M-8076/2 Carcinoma, squamous cell, in situ with questionable stromal invasion (C53._)	*Delete:* (C53._)
124	2	23	x	x	M-8072/3 Carcinoma, squamous cell, large cell, nonkeratinizing	*Add at end of line:* , NOS
124	2	25	x	x	M-8076/3 Carcinoma, squamous cell microinvasive (C53._)	*Delete:* (C53._)
124	2	52	x	x	*In space above* M-8211/3 Carcinoma, tubular	*Insert new term even with 'tubular':* M-8102/3 trichilemmal (C44._)

PG	COL	LINE	HC	SC	TERM	ACTION
128	2	7	x		*In space after* M-9242/3 Clear cell (type), chondrosarcoma (C40._, C41._)	*Insert new term even with 'chondrosarcoma':* M-8313/3 cystadenocarcinofibroma (C56.9)
128	2	11	x		M-8313/3 Clear cell (type), cystadenocarcinofibroma (C56.9)	*Delete line (out of sequence)*
137	2	33	x	x	M-9383/1 Ependymoma-subependymoma, mixed	*Add at end of line:* (C71._)
137	2	46	x	x	M-8076/2 Epidermoid carcinoma, in situ with questionable stromal invasion (C53._)	*Delete:* (C53._)
141	2	49	x	x	*In space after* M-9041/3 Fibrous synovial sarcoma, monophasic	*Add term even with 'synovial':* tissue (see next page)
144	1	40	x	x	M-8043/3 Fusiform cell, small cell carcinoma (C34._)	*Delete:* (C34._)
145	1	20	x	x	M-9064/2 Germ cell, intratubular malignant (C62._)	*Delete line*
145	1	25	x	x	*In space above* M-9064/3 Germinoma	*Insert new term even with 'germinoma':* M-9064/2 Germ cells, intratubular malignant (C62._)
147	1	7	x		M-9073/1 Gonadoblastoma	*Delete indent (align under'Gonadal' above)*
147	1	8	x		M-9073/1 Gonocytoma	*Delete indent (align under'Gonadal' above)*
151	2	11	x	x	M-------- Immunoblastic lymphadenopathy (see SNOMED)	*Replace code with:* M-9767/1 *Delete:* (see SNOMED) *Add at end of line:* (IBL) [obs]
151	2	17	x		*In space above* M-9760/3 Immunoproliferative disease, NOS	*Insert new term even with 'disease':* M-9766/1 angiocentric lesion
151	2	56	x	x	M-8076/2 In situ, epidermoid carcinoma with questionable stromal invasion (C53._)	*Delete:* (C53._)
153	1	41	x	x	M-8044/3 Intermediate cell, small cell carcinoma (C34._)	*Delete:* (C34._)
153	2	38	x	x	*In space after* M-8504/2 Intracystic carcinoma, noninfiltrating	*Insert new term even with 'carcinoma':* M-8504/3 carcinoma, papillary
153	2	40	x	x	*In space after* M-8504/0 Intracystic papillary adenoma	*Insert new term even with 'papillary':* M-8504/3 papillary carcinoma
154	2	55	x	x	M-8150/0 Islet cell tumor, benign	*Add at end of line:* (C25._)

PG	COL	LINE	HC	SC	TERM	ACTION
155	2	17	x		M-9160/0 Juvenile angiofibroma (C11._)	*Delete:* (C11._)
156	2	44	x	x	M-8072/3 Large cell carcinoma, squamous cell, nonkeratinizing	*Add at end of line:* , NOS
166	1	24	x	x	M-9767/1 Lymphadenopathy, angioimmunoblastic	*Add at end of line:* (AIL)
166	1	28	x	x	M-------- Lymphadenopathy, immunoblastic (see SNOMED)	*Replace code with:* M-9767/1 *Delete:* (see SNOMED) *Add at end of line:* (IBL) [obs]
175	2	24	x	x	M-9383/1 Mixed ependymoma-subependymoma	*Add at end of line:* (C71._)
175	2	34	x		M-8180/3 Mixed hepatocellular and bile duct carcinoma	*Add at end of line:* (C22.0)
176	1	17	x		M-9383/1 Mixed subependymoma-ependymoma	*Add at end of line:* (C71._)
182	2	7	x	x	M-8072/3 Nonkeratinizing, squamous cell carcinoma, large cell	*Add at end of line:* , NOS
185	2	23	x	x	*In space after* M-8052/2 Papillary, carcinoma, in situ, squamous cell	*Insert new term even with 'in situ':* M-8504/3 intracystic
186	1	9	x	x	*In space after* M-8405/0 Papillary, hidradenoma (C44._)	*Insert TWO new terms even with 'hidradenoma':* M-8504/3 intracystic adenocarcinoma M-8504/3 intracystic carcinoma
190	1	46	x		M-9733/3 Plasma cell leukemia (C42.1)	*Delete line (listed under plasma cell, above)*
193	2	19	x	x	M-8076/2 Questionable stromal invasion, epidermoid carcinoma in situ with (C53._)	*Delete:* (C53._)
193	2	21	x	x	M-8076/2 Questionable stromal invasion, squamous cell carcinoma in situ with (C53._)	*Delete:* (C53._)
195	1	32	x		*In space after* M-8900/3 Rhabdomyosarcoma, NOS	*Insert new term even with 'NOS'* M-8901/3 adult type
195	1	40	x		M-8901/3 Rhabdomyosarcoma, adult type	*Delete line (out of sequence)*
197	2	22	x		M-8350/3 Sclerosing, tumor, nonencapsulated sclerosing (C73.9)	*Delete 'sclerosing'*
201	1	6	x	x	M-8043/3 Small cell, carcinoma, fusiform cell (C34._)	*Delete:* (C34._)

PG	COL	LINE	HC	SC	TERM	ACTION
201	1	7	x	x	M-8044/3 Small cell, carcinoma, intermediate cell (C34._)	*Delete:* (C34._)
203	1	13	x		M-9423/3 Spongioblastoma, primitive polar 71._) [obs]	*Add opening parenthesis:* (C71._)
203	1	40	x	x	M-8076/2 Squamous cell, carcinoma, in situ with questionable stromal invasion (C53._)	*Delete:* (C53._)
203	1	45	x	x	M-8072/3 Squamous cell, carcinoma, large cell, nonkeratinizing	*Add at end of line:* , NOS
203	1	47	x	x	M-8076/3 Squamous cell, carcinoma, microinvasive (C53._)	*Delete:* (C53._)
205	1	7	x	x	M-9383/1 Subependymoma-ependymoma, mixed	*Add at end of line:* (C71._)
211	2	56	x		M-9260/3 Tumor, Ewing tumor (C40._, C41._)	*Delete 'tumor'*
212	2	24	x		M-8150/0 Tumor, islet cell, benign	*Add at end of line:* (C25._)
214	1	37	x		M-8641/0 Tumor, Sertoli cell, lipid-rich	*Add at end of line:* (C56.9)
222	1	29	x	x	8825/1 Inflammatory myofibroblastic tumor	*Delete code (synonym of previous term)*
225	1	44	x		9871/3 Acute myeloid leukemia with abnormal marrow eosinophils (includes all variants)	*Add ** after code*
225	1	50	x		9872/3 Acute myeloid leukemia, minimal differentiation	*Add ** after code*
225	1	52	x		9873/3 Acute myeloid leukemia without maturation	*Add ** after code*
225	1	after last line	x			*Add note:* ** Code used in United States and Canada (1998-2000)
226	1	1	x		9874/3 Acute myeloid leukemia with maturation	*Add ** after code*
226	1	after last line	x			*Add note:* ** Code used in United States and Canada (1998-2000)
237	1	31	x		9865/3** Acute myeloid leukemia with maturation	*Delete line (correct code and term on page 226)*
237	1	32	x		9865/3** FAB M2, NOS	*Delete line (correct code and term on page 226)*
237	1	34	x		9869/3** Acute myeloblastic leukemia	*Delete line (correct code and term on page 225)*
237	1	35	x		9869/3** FAB M0	*Delete line (correct code and term on page 225)*

Additional ICD-O-3 Clarifications

1. **[obs]** The use of the [obs] is not well described in ICD-O-3. This descriptor is intended to discourage the use of such a term for a new diagnosis when better diagnostic terms are available. If a term marked [obs] is diagnosed, it may certainly be coded, although it is likely that a more current term is available. If the [obs] term is a reportable malignancy (typically /2 and /3 behavior codes), DO include it in the registry even though the terminology is out of date. Furthermore, [obs] serves 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 argentaffinoma [obs] which is now described as carcinoid tumor with additional codes for several variants. 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 that had specific codes in ICD-O-2 have been moved to the 'Not Otherwise Specified' category for the disease.

2. **No Rule "I"** There is no "Rule I" in ICD-O-3 and this was done intentionally. The rules in ICD-O-2 were numeric. The rules in ICD-O-3 are alphabetic. The editors of ICD-O-3 felt it necessary to omit Rule I from ICD-O-3 in an attempt to avoid any possible confusion between the 1 (one) and I (the letter 'i'), as in "Rule 1 (one)" in ICD-O-2 and "Rule I (the letter 'i')" in ICD-O-3.

3. **Rule D: coding extranodal lymphomas** Due to a printer's error, the wording of Rule D on page 20 of the hardcover version is not the same as the wording of Rule D on page 26 in the hardcover version and Rule D on pages 20 and 26 in the softcover version. As indicated in Table 2 of this document, please replace the text of Rule D on page 20 in the hardcover version with the text of Rule D on page 26. The softcover version is correct as printed. The difference occurred in the late stages of ICD-O-3 editorial review when the following statement was added to the rule: "If no site is indicated for a lymphoma and it is suspected to be extranodal, code to C80.9 (unknown primary site)." This statement was added to reduce the number of extranodal lymphomas coded to lymph node, NOS (C77.9) when the site of origin is unknown, and it will apply only in a very limited set of circumstances. In most cases, the site of origin of an extranodal lymphoma is known, and the topography should be coded to that site, such as a primary lymphoma of the stomach. However, if the site of origin is unclear and there is no evidence of lymphoma in lymph nodes, it would be appropriate to code the lymphoma to unknown primary site. For example, if the patient has bulky disease in both the lung and paraspinal soft tissues (without lymph nodes involved), it may not be possible to determine which location is the site of origin; therefore, coding to C80.9 is correct. Another example would be a death certificate only case or one diagnosed at a different facility and reported as an "extranodal" lymphoma but the site of origin is not specified. This case is also appropriately coded to C80.9. Bear in mind that there is no change in coding guidelines for lymphomas arising in lymph nodes or in lymphoid tissues. It is understood that coding lymphomas to an unknown primary is a new concept; computer edits are being revised to accommodate them.

4. **Using a grade designation to assign 6th digit differentiation** In some instances, the term "grade" does not imply differentiation and should not be used to code the 6th digit of the morphology code. For example, in describing some diseases, 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 that is coded in the 6th digit of the ICD-O morphology code. It is important to recognize when the term "grade" refers to category and when it refers to biologic activity. For example, the grades of nodular sclerosing Hodgkin lymphoma and follicular lymphoma are actually types or categories of these diseases. The 6th digit should NOT be coded as grade 1, 2 or 3 for these cases. However, a poorly-differentiated lymphocytic lymphoma or a B-cell or T-cell lymphoma should be coded in the 6th digit of the morphology code. Similarly, vaginal intraepithelial neoplasia, grade III (VAIN III) is actually the highest category of dysplasia (according to the Bethesda system) for a non-invasive lesion. Grades associated with intraepithelial neoplasias should NOT be used to code the morphology 6th digit. However, other terms described as high grade or low grade as part of the diagnostic term, such as low grade endometrial stromal sarcoma and high grade surface osteosarcoma, may be used to code the 6th digit of the morphology code.

5. **Assigning 6th digit immunophenotype** Sixth digit codes for T-cell, B-cell, and NK-cell phenotyping of lymphomas and leukemias should be based on the diagnosis as specifically stated in the pathology report. Sixth digit phenotype codes should not be used when T- or B- cell is implied from the boldface header in the morphology numeric list. In other words, if no T- or B-cell designation is provided in the pathology or laboratory report, do NOT code the T- or B- cell designation based on the boldface header in ICD-O-3. For example, a diffuse large B-cell lymphoma would be coded to 9680/36; a diffuse centroblastic lymphoma would be coded to 9680/39. When cases are analyzed, they can be grouped by cell line as stated in the category headings in the lymphoma and leukemia sections of the morphology numeric list.

6. **Assigning topography for hematopoietic diseases** According to the medical understanding on which the World Health Organization Classification of Hematopoietic Neoplasms is based, 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 (BCCLL/SLL) as a single entity, the same disease at different stages. The hemato-pathologists 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, some single disease entities are listed in two different categories and cross-referenced with the notation (see also M-9----). The topographic or primary site code for a diagnosis such as BCCLL/SLL 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 and assign the leukemia morphology code. If the diagnosis is made on any other tissue (typically lymph nodes, lymphatic structures, breast, and stomach), code to the tissue involved and assign the lymphoma morphology. If the diagnosis is made on both blood or bone marrow and a tissue biopsy, code the tissue involved and assign the lymphoma morphology. The sequence of the biopsies (whether the blood/bone marrow biopsy is done before the tissue biopsy or vice versa) is not a factor in deciding which primary site and morphology code to use. For purposes of analysis according to the WHO Classification, cases from both morphology codes should be aggregated.

7. **"Code to the higher morphology code"** The general ICD-O-3 guideline to use the numerically higher morphology code if the diagnosis of a single tumor includes two modifying adjectives with different code numbers (Rule K) does not apply to separate or independent solid tumors nor to the hematopoietic diseases (M-9590-9989) in general. For the hematopoietic diseases, code to the more specific morphology, if that can be determined, which may not be the numerically higher code number. For example, if the facility pathology report states "diffuse large B-cell lymphoma" (M-9680/3) and a consultant reports the same tissue to be "mantle cell lymphoma" (M-9673/3), code the case to M-9673/3. The primary term for M-9680/3 includes the term NOS (not otherwise specified) and the code contains 27 synonyms; thus it can be considered a non-specific diagnosis. On the other hand, the primary term for M-9673/3 does not include the term NOS and may therefore be considered more specific. When in doubt which code to use, consult a medical advisor or pathologist.

8. **Combination breast histology codes** For breast cancer cases with multiple subtypes, two new codes have been included in ICD-O-3. Use code 8523 when there is a diagnosis of duct carcinoma mixed with another carcinoma or more than one subtype of ductal carcinoma, such as ductal carcinoma with elements of cribriform, mucinous and lobular carcinoma or duct carcinoma mixed with mucinous carcinoma. The same principle applies for 8524 when one of the histologies is lobular carcinoma. Although there are no equivalent terms specifically listed under the primary term for 8524/3, such as those shown for 8522/3 and 8523/3, the "other types of carcinoma" with which the lobular carcinoma is mixed can include histologies such as mucinous, tubular, cribriform and/or solid. Bear in mind that if all parts of the tumor are in situ, the behavior code should be /2 (in situ). If any part of the tumor is invasive, the behavior must be /3 (invasive).