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Reviewing the ICD-10 classification of haematological neoplasms on its way to ICD-11

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Abstract: Reviewing the ICD-10 classification of haematological neoplasms on its way to ICD-11

Under the leadership of the German WHO-FIC Collaborating Centre the section for the haematological neoplasms was reviewed. This paper will explain methods and results.

At the last WHO-FIC Network meeting in Reykjavik areas that need to be reviewed more systematically on the way to ICD-11 have been identified.

A two-step approach for the haematological neoplasm was recommended: the way towards a restructured leukemia and lymphoma section in ICD-11 should be paved by a moderate update of ICD-10 in order to bring this section in line with ICD-O-3 and the current state of science and to achieve a smoother transition to ICD-11 without major breaks in health statistics.

Due to great discrepancies between classifications in use and the current state of science clinicians and pathologists seek for updating ICD-10 in accordance with the Update Reference Committee (URC) and the WHO-FIC Network as soon as possible.

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Introduction

At the WHO Family of International Classifications (WHO-FIC) Network Meeting 2004 (Reykjavik, Iceland) it was agreed that the German centre, as others, takes part in reviewing ICD-10 in distinct areas which were identified as crucial and representative for other sections [1].

As areas for reviewing the evidence of ICD-10 and starting the process cystic fibrosis, diabetes, lymphomas, external causes and mental health were identified.

The German Collaborating Centre (CC) took responsibility for reviewing the section of leukaemias and lymphomas.

Main issues regarding the classification of haematological neoplasm in ICD-10 are:

- differences between ICD-10 and ICD-O-3 concerning the structure
- differences between ICD-10 and ICD-O-3 concerning the content
- differences between ICD-10, ICD-O-3 and the current state of science
- limitations due to the updating of ICD-10

This document will explain methods and results.

Working group

A board of national experts in haemato-oncology was formed of pathologists, clinicians, DRG experts, specialized practitioners and statisticians.

Following experts joined the board:

Prof. Dr. med. Stein / University of Berlin Charité - Campus Benjamin Franklin / coauthor of blue book [2]

PD Dr. med. Krause / University of Regensburg / German Society of Hematology and Oncology

Prof. Dr. med. Berthold / University of Cologne / Society of Pediatric Oncology and Hematology

Prof. Dr. med. Wittekind / University of Leipzig / Pathologist

Prof. Dr. med. Schlake / President of the Association of German Pathologists

Dr. med. Bertz / Robert Koch Institut / Central Cancer Surveillance Program - Main Cancer Registry

Dr. med. Christaras / University of Düsseldorf / Society of Pediatric Oncology and Hematology

Dr. med. Maywald / Competence Network Acute and Chronic Leukemia

Dr. med. Aly / Network of Specialized General Practitioners in Oncology, Competence Network Malignant Lymphoma

Robert Jakob / WHO Headquarters Geneva

Dr. med. Michael Schopen / DIMDI / Medical Classification Section

Ulrich Vogel / DIMDI / Medical Classification Section

Method

At the last WHO-FIC meeting in Reykjavik a two-step approach was recommended for ICD-11: the way towards a restructured leukemia and lymphoma section in ICD-11 should be paved by a moderate update of ICD-10:

Over the years new entities were introduced and old entities were removed from the classification of leukaemia and lymphoma described in the blue book [2] and ICD-O-3. These changes should be incorporated into ICD-10 without structural breaks.

In a first session the limitations, rules and needs for updating of ICD-10 were specified. It was emphasized that no codes should be reassigned and that no fifth digit codes of the German Modification of ICD-10 can be introduced. The structural changes should be as small as possible and mainly be achieved by deletion of old and insertion of new categories and by insertion and deletion of inclusion or exclusion notes.

One proposal from the clinicians and one from the pathologists were merged to one first draft version. Afterwards it was revised several times.

Although some proposals of the clinicians using the German Modification of ICD-10 were also driven by the needs of Diagnoses Related Groups (DRG) and reimbursement, we made sure to stay in line with the needs of the official WHO version.

Thus the content of the Blue Book for Lymphomas and Leukaemias [2], the structures of ICD-O-3 and the existing ICD-10 could be respected.

Bringing this section in line with ICD-O-3 and the current state of science will achieve a smoother transition to ICD-11 as crosswalks at the level of entities will prevent major breaks in health statistics.

Results

The significant entities in haemato-oncology at the current state of science could be placed in the existing part of ICD-10. Because of the restrictions imposed by existing 3- and 4-character categories, the main focus was set on the correct placement of the content. The overall view of modifications in figure 1 gives a first impression of the approach and identifies “hot spots”. Numbers of changes made are as follows (figure 2): Of 20 3-character categories 7 (35%) were modified, 2 new categories were introduced and none were removed. Overall percentage of changes of relevant 3-character categories was 45 (%). Of 126 4-character subcategories 57 (45%) were modified, 31 (24%) new subcategories were introduced and 12 (9%) were removed from the classification. Overall percentage of changes of relevant subcategories was 79 (%).

This number of changes might seem overwhelming at first sight.

Indeed changes of content at a 3 character category level reflect the new knowledge in the field. They do not always affect the content of a 3 character category. A good example would be the change of the title of C84. The old as well as the new title reflect the same content only the text had to be adapted to new knowledge. As the number of available 4 character subcategories of C84 was not sufficient to list all relevant entities, the new C86 was created. The entities listed under this new code were comprised in C84.5 in the past (C84.old=C84.new+C86.new).

A price for the integration of all entities into the somewhat outdated system of the ICD-10 is the separation of closely related diagnoses into different 3-character categories on one side and grouping of relatively unrelated entities into one 3-character category on the other side.

C88.1, Alpha heavy chain disease, is now known to mean exactly the same as C88.3, Immunoproliferative small intestinal disease (Mediterranean disease). The name of this disease was different according to the region the patient was diagnosed. The frequencies for C88.3 in the countries concerned apparently have decreased significantly, nevertheless it was decided to keep this code

instead of the C88.1. This would allow the monitoring of this disease without any change for the relevant countries. The Alpha heavy chain disease (C88.1) in the non mediterranean countries is a rare disease and the disappearing of the code would be insignificant and the disease can still be identified with the C88.3.

The changes between the old and the new C83 seem huge (see appendix). Indeed many of them are just clarifications and allow the identification of new lymphoma entities, hidden in the old structure within other terms. The old and the new C83 will code nearly the same group of patients.

For use at international level the stability of the 3 character categories in the sophisticated and rapidly changing field of lymphomas and leukaemias may be sufficient. For reimbursement more detail is required and has to be up to date to reflect the treatment (costs) as well as the prognosis. For scientific purposes ICD-10 can hardly provide the full detail that ICD-O already does.

Seeking for international agreement first results have already been shared with the International Agency for Research on Cancer (IARC) by WHO. An official statement from IARC is pending. Still, we do think results can already support the updating process of ICD-10 in short- and mid-term.

The preliminary results are provided in two separate documents. One shows the restructured and updated tabular list, the other one presents a synopsis of the current ICD-10 and the new proposal (see attached documents).

Preliminary crosswalks are under construction and will be available in October 2005. As a second step clinicians and DRG experts will prepare DRG-mappings of this new classification in cooperation with the German DRG institute (InEK). First results can be expected in late 2005.

Entries in both the Alphabetic Index and the Morphology of Neoplasms will need an appropriate review as well. Due to the amount of modifications this challenge should be taken into account, too. However, this work should start after the endorsement of the changes to ICD-10.

From today's perspective there is a broad range of institutions, medical societies and legal authorities in German supporting an update of the German Modification of ICD-10.

When international feedback to this proposal is available a consensus proposal will be submitted to the URC and the WHO-FIC Network for implementation in ICD-10 as a first step. Afterwards a proposal for structure and content of ICD-11 can be developed.

References

[1] Meeting Report of WHO Family of International Classifications Network Meeting 2004, Reykjavik (Finland)

[2] WHO Classification of Tumours, Volume 3 [IARC WHO Classification of Tumours, No 3] Jaffe, E.S., Harris. N.L., Stein, H., Vardiman, J.W., 2001

Annex

Parts of the current version of "**haematological neoplasms"** in ICD-10 have been deleted. New parts have been added. In the following tables and figures those differences are compared. Not all parts have been changed, but in order to have a clear representation the whole section has been displayed.

The following documents are provided separately:

1. Tabular list with new proposals

   (leukaemia_lymphoma_paper_annexdoc1_tokyo_20050927.rtf)

2. Synopsis of tabular lists of WHO Version of ICD-10 2005 and new proposal

   (leukaemia_lymphoma_paper_annexdoc2_tokyo_20050927.rtf)

Figure 1: Overall view of modifications to 3- and 4-character categories

Categories		not modified	modified	new	deleted
C81	Hodgkin lymphoma		X		
C81.0	Lymphocyte-rich classical Hodgkin lymphoma		X		
C81.1	Nodular sclerotic classic Hodgkin lymphoma		X		
C81.2	Mixed cellularity classical Hodgkin lymphoma		X		
C81.3	Lymphocyte depleted classical Hodgkin lymphoma		X		
C81.4	Nodular lymphocyte predominant Hodgkin lymphoma			X	
C81.7	Other classical Hodgkin lymphoma		X		
C81.9	Hodgkin lymphoma, unspecified		X		
C82	Follicular lymphoma		X		
C82.0	Follicular lymphoma grade I (small cell, cleaved nucleus)		X		
C82.1	Follicular lymphoma grade II (mixed small and large cell, cleaved nucleus)		X		
C82.2	Follicular lymphoma grade III, unspecified		X		
C82.3	Follicular lymphoma grade IIIa			X	
C82.4	Follicular lymphoma grade IIIb			X	
C82.5	Diffuse follicle center lymphoma		X		
C82.6	Cutaneous follicle center lymphoma			X	
C82.7	Other types of follicular lymphoma		X		
C82.9	Follicular lymphoma, unspecified		X		
C83	Non-follicular lymphoma		X		
C83.0	Small cell B-cell lymphoma		X		
C83.1	Mantle cell lymphoma		X		
~~C83.2~~	~~Mixed small and large cell (diffuse)~~				X
C83.3	Diffuse large B-cell lymphoma		X		
~~C83.4~~	~~Immunoblastic (diffuse)~~				X
C83.5	Lymphoblastic lymphoma		X		
~~C83.6~~	~~Undifferentiated (diffuse)~~				X
C83.7	Burkitt's lymphoma		X		
C83.8	Other non-follicular lymphoma		X		
C83.9	Non-follicular (diffuse) non-Hodgkin lymphoma, unspecified		X		
C84	Peripheral T/NK-cell lymphomas		X		
C84.0	Mycosis fungoides	X			
C84.1	Sézary syndrome		X		
C84.2	T-zone lymphoma	X			
C84.3	Lymphoepithelioid lymphoma	X			
C84.4	Unspecified peripheral T-cell lymphoma		X		
C84.5	Other T/NK-cell lymphomas		X		
C84.6	Anaplastic large cell lymphoma, ALK-positive			X	
C84.7	Anaplastic large cell lymphoma, ALK-negative			X	
C84.9	Peripheral T/NK-cell lymphoma, unspecified			X	
C85	Other and unspecified types of non-Hodgkin's lymphoma	X			
~~C85.0~~	~~Lymphosarcoma~~				X
C85.1	B-cell lymphoma, unspecified	X			
C85.2	Mediastinal [thymic] large B-cell lymphoma			X	
~~C85.7~~	~~Other specified types of non-Hodgkin's lymphoma~~				X
C85.9	Non-Hodgkin's lymphoma, unspecified	X			

Categories		not modified	modified	new	deleted
C86	Other specified types of T/NK-cell lymphoma			X	
C86.0	Extranodal NK-cell lymphoma, nasal type			X	
C86.1	Hepatosplenic T-cell lymphoma			X	
C86.2	Enteropathy-type T-cell lymphoma			X	
C86.3	Subcutaneous panniculitis-like T-cell lymphoma			X	
C86.4	Blastic NK-cell lymphoma			X	
C86.5	Angioimmunoblastic T-cell lymphoma			X	
C86.6	Primary cutaneous CD30-positive T-cell proliferations			X	
C88	Other B-cell lymphoma [malignant immunoproliferative diseases]		X		
C88.0	Waldenström's macroglobulinaemia		X		
~~C88.1~~	~~Alpha heavy chain disease~~				X
C88.2	Gamma heavy chain disease		X		
C88.3	Immunoproliferative small intestinal disease [alpha heavy chain disease]		X		
C88.4	Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue [MALT-lymphoma]			X	
C88.7	Other malignant immunoproliferative diseases	X			
C88.9	Malignant immunoproliferative disease, unspecified	X			
C90	Plasmacytoma and malignant plasma cell neoplasms		X		
C90.0	Multiple myeloma		X		
C90.1	Plasma cell leukaemia	X			
C90.2	Solitary plasmacytoma		X		
C91	Lymphoid leukaemia	X			
C91.0	Acute lymphoblastic leukaemia		X		
C91.1	Chronic lymphocytic leukaemia of B-cell type		X		
~~C91.2~~	~~Subacute lymphocytic leukaemia~~				X
C91.3	Prolymphocytic leukaemia of B-cell type		X		
C91.4	Hairy cell leukaemia	X			
C91.5	Adult T-cell lymphoma/leukaemia (HTLV-1-associated)		X		
C91.6	Prolymphocytic leukaemia of T-cell type			X	
C91.7	Other lymphoid leukaemia		X		
C91.9	Lymphoid leukaemia, unspecified	X			
C92	Myeloid leukaemia		X		
C92.0	Acute myeloblastic leukaemia [AML]		X		
C92.1	Chronic myeloid leukaemia [CML], BCR/ABL-positive		X		
C92.2	Atypical chronic myeloid leukaemia, BCR/ABL-negative		X		
C92.3	Myeloid sarcoma		X		
C92.4	Acute promyelocytic leukaemia [PML]		X		
C92.5	Acute myelomonocytic leukaemia		X		
C92.6	Acute myeloid leukaemia with 11q23-abnormity			X	
C92.7	Other myeloid leukaemia		X		
C92.8	Acute myeloid leukaemia with multilineage dysplasia			X	
C92.9	Myeloid leukaemia, unspecified		X		
C93	Monocytic leukaemia	X			
C93.0	Acute monoblastic and monocytic leukaemia		X		
C93.1	Chronic myelomonocytic leukaemia		X		
C93.2	Juvenile myelomonocytic leukaemia		X		
C93.7	Other monocytic leukaemia	X			
C93.9	Monocytic leukaemia, unspecified	X			

Categories		not modified	modified	new	deleted
C94	Other leukaemias of specified cell type	X			
C94.0	Acute erythroid leukaemia		X		
~~C94.1~~	~~Chronic erythraemia~~				X
C94.2	Acute megakaryoblastic leukaemia		X		
C94.3	Mast cell leukaemia	X			
C94.4	Acute panmyelosis with myelofibrosis		X		
~~C94.5~~	~~Acute myelofibrosis~~				X
C94.6	Myelodysplastic / myeloprolifarative disease, not classifiable			X	
C94.7	Other specified leukaemias		X		
C95	Leukaemia of unspecified cell type	X			
C95.0	Acute leukaemia of unspecified cell type		X		
C95.1	Chronic leukaemia of unspecified cell type		X		
C95.2	Subacute leukaemia of unspecified cell type	X			
C95.7	Other leukaemia of unspecified cell type	X			
C95.9	Leukaemia, unspecified	X			
C96	Other and unspecified malignant neoplasms of lymphoid, haematopoietic and related tissue	X			
C96.0	Multifocal and multisystemic (disseminated) Langerhans-cell histiocytosis [Letterer-Siwe disease]		X		
C96.1	Histiocytic sarcoma		X		
C96.2	Malignant mast cell tumour		X		
~~C96.3~~	~~True histiocytic lymphoma~~				X
C96.4	Sarcoma of dendritic cells (accessory cells)			X	
C96.5	Multifocal and unisystemic Langerhans-cell Histiocytosis			X	
C96.6	Unifocal Langerhans-cell Histiocytosis			X	
C96.7	Other specified malignant neoplasms of lymphoid, haematopoietic and related tissue	X			
C96.9	Malignant neoplasm of lymphoid, haematopoietic and related tissue, unspecified	X			
C97	Malignant neoplasms of independent (primary) multiple sites	X			
D45	Polycythaemia vera	X			
D46	Myelodysplastic syndromes	X			
D46.0	Refractory anaemia without ringed sideroblasts, so stated		X		
D46.1	Refractory anaemia with ringed sideroblasts, so stated		X		
D46.2	Refractory anaemia with excess of blasts		X		
~~D46.3~~	~~Refractory anaemia with excess of blasts with transformation~~				X
D46.4	Refractory anaemia, unspecified	X			
D46.5	Refractory anaemia with multi-lineage dysplasia			X	
D46.6	Myelodysplastic syndrome with isolated del(5q) chromosomal abnormality			X	
D46.7	Other myelodysplastic syndromes		X		
D46.9	Myelodysplastic syndrome, unspecified	X			
D47	Other neoplasms of uncertain or unknown behaviour of lymphoid, haematopoietic and related tissue	X			
D47.0	Histiocytic and mast cell tumours of uncertain and unknown behaviour		X		
D47.1	Chronic myeloproliferative disease	X			
D47.2	Monoclonal gammopathy	X			
D47.3	Essential (haemorrhagic) thrombocythaemia	X			
D47.4	Osteomyelofibrosis			X	
D47.5	Chronic eosinophilic leukaemia [hypereosinophilic syndrome]			X	
D47.7	Other specified neoplasms of uncertain or unknown behaviour of lymphoid, haematopoietic and related tissue	X			
D47.9	Neoplasm of uncertain or unknown behaviour of lymphoid, haematopoietic and related tissue, unspecified	X			

Categories		not modified	modified	new	deleted
D75	Other diseases of blood and blood-forming organs	X			
D75.0	Familial erythrocytosis	X			
D75.1	Secondary Polycythaemia	X			
~~D75.2~~	~~Essential thrombocytosis~~				X
D75.8	Other specified diseases of blood and blood-forming organs	X			
D75.9	Disease of blood and blood-forming organs, unspecified	X			
D76	Specified diseases with participation of lymphoreticulary and reticulohistiocytic tissue			X	
D76.1	Haemophagocytic lymphohistiocytosis			X	
D76.2	Haemophagocytic syndrome, infection-associated			X	
D76.3	Other histiocytosis syndromes			X	
	No. of categories (147)	38	64	33	12
	Percentage (100%)	26%	44%	22%	8%

Figure 2: Number of revised 3- and 4-character categories (figure 1)

	not modified	modified	new	deleted	total
No. of 3-char. categories	11	7	2	0	20
	55 %	35 %	10 %	0 %	100 %
	55 %		45 %		100 %
No. of 4-char. categories	27	57	31	12	127
	21 %	45 %	24 %	9 %	100 %
	21 %		79 %		100 %