APPLICATION OF THE INTERNATIONAL CLASSIFICATION OF DISEASES TO NEUROLOGY

ICD-NA

(Published for trial purposes)

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PREFACE

On the initiative of the World Health Organization, a consultation was convened in 1984 to consider the classification of diseases of the nervous system in relation to the Ninth Revision of the International Classification of Diseases (1975) and the forthcoming Tenth Revision. The need for a more detailed classification for use in morbidity statistics, hospital record indexing and research in the field of neurology was recognized. The Application of the International Classification of Diseases to Neurology provides such a classification. The World Federation of Neurology, its Research Committee, and individual research group collaborated in its preparation.

This edition is published mainly for trial purposes; it needs to be tested in various areas of neurology to determine its usefulness. Comments on the application of the classification are welcome and should be sent to:
Neurosciences Programme
Division of Mental Health
World Health Organization
1211 Geneva 27
Switzerland.

ACKNOWLEDGEMENTS

INTRODUCTION

When any substantial volume of data has to be recorded, a system of classification and coding is necessary. Coding is especially important if the data are to be retrieved or analysed by automatic means.

The Application of the International Classification of Diseases to Neurology (ICD-NA) is intended to provide a basis for such classification and coding. Within the framework of the Ninth Revision of the International Classification of Diseases (ICD-9), the ICD-NA aims to provide a convenient coding method for use by those concerned with disorders of the nervous system.

ICD-NA is derived directly from the Ninth Revision of the ICD[1,2] and covers diseases and conditions that occur in, have manifestations in, or have associations with the nervous system and adjacent structures.

Classifications and terminology used in the ICD (Ninth Revision) have been expanded. In general, data collected with ICD-NA can be assembled into ICD categories by simple addition. It is recommended that the ICD-NA be used in conjunction with the Ninth Revision of the ICD. The reasons for not using the ICD alone are:

(1) the diseases and conditions of interest to neurologists, neurosurgeons, and neuroscientists are insufficiently subdivided, and

(2) such diseases and conditions are scattered throughout the ICD, which imposes difficulties in its use by neurologists and neurosurgeons.

The aims of the ICD-NA are:

(1) to focus the attention of specialists in neurology on the desirability of a detailed diagnosis for each patient, using a comprehensive and consistent classification of neurological diseases and of neurological manifestations of other diseases.

(2) to provide an improved standard recording system for neurological diseases and conditions.

(3) to make possible the collection of epidemiological data and comparisons of the prevalence of neurological diseases at both national and

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[1] WORLD HEALTH ORGANIZATION. *Manual of the International Statistical Classification of Diseases, Injuries, and Causes of Death* (ninth revision), Vol. 1, Geneva, 1977.

[2] WORLD HEALTH ORGANIZATION. *Manual of the International Statistical Classification of Diseases, Injuries, and Causes of Death* (ninth revision), Vol. 2, Geneva, 1978.

international level. It is hoped that the system will also facilitate the collection of epidemiological data on the rarer neurological diseases.

In addition, the classification should serve to promote investigations of the distribution of neurological disease and the identification of risk factors. Such information is urgently needed to support national programmes of prevention and control.

It is hoped that the ICD-NA will be of value to a great variety of users, ranging from governments concerned with the collection of statistical data under relatively few main headings to individual physicians requiring a convenient tool for the indexing of their clinical and teaching material. The ICD-NA is capable of contraction to a few broad categories, or of further expansion in areas in which the user may have a special interest. Finally, the ICD-NA provides a method of classification that facilitates international collaboration and exchange of information.

MANUAL OF THE INTERNATIONAL STATISTICAL CLASSIFICATION OF DISEASES, INJURIES, AND CAUSES OF DEATH (ICD)

Both the reader and the user are referred to the Ninth Revision of the ICD for general principles, historical and recent background, and description of the ICD classification. The following description presents only those features essential to use of the ICD-NA.

The ICD is a systematic classification of diseases, subject to agreement by governments. It is widely used for national mortality and morbidity statistics and is revised decennially. The Ninth Revision came into effect on 1 January 1979, and consists of two volumes. The first includes an explanatory text and a tabular (numerical) presentation of the classification. The second lists all items of the classification alphabetically. The taxonomic philosophy of the ICD is somewhat eclectic, as no strictly systematic classification is really practicable, because of different national views as to disease classification and terminology. The classification scheme of the ICD has been followed for the ICD-NA. The ICD also includes a coded nomenclature of the morphology of neoplasms, an extract of which is included in the present volume.

Not every condition receives a particular rubric or number, but there is a category to which every condition can be referred, and this has been achieved by the method of selective grouping. The principles of determining what conditions should be specified as definite categories are based on frequency, importance, and clarity of characterization of the condition.

A decimal system of numbering has been adopted in which the detailed categories of the classification are designated by 3-digit numbers.

In many instances, the first 2 digits of the 3-digit number designate important or summary groups that are significant. The third digit divides each group into categories that represent specific disease entities or a classification of the disease or condition according to some significant axis, such as anatomical site. Further, the detailed or 3-digit categories have not been numbered consecutively, but numbers have been omitted in order that the summary character of the first 2 digits could be preserved wherever it is meaningful. No additional 3-digit categories may be introduced in the classification, except when the list is revised by international agreement.

The ICD also contains a fourth digit designed for more comprehensive studies of the causes of illness and disability. An attempt has been made to show most of the diagnostic terms given in the standard or official nomenclatures, as well as terms commonly used in different countries; these terms have been called "inclusion terms". Where there is a reasonable risk that a condition will be wrongly classified, cross-reference to relevant categories is achieved by "exclusion terms". The last 2 numbers of the fourth digit (.8 and .9) very often carry the connotation "other" and "unspecified", respectively. "NOS" is an abbreviation for "not otherwise specified" and is virtually the equivalent of "unspecified" or "unqualified".

THE ICD-NA

The ICD-NA, like the ICD, has a tabular (numerical) section and a comprehensive alphabetical index. Liberal use has been made of inclusion and exclusion terms in the tabular section, the latter being provided with classification numbers, so that the user will have as much assistance as possible in finding the correct category for any condition diagnosed. In addition, certain notes of classification and cross-references (given in round brackets) have been added to facilitate use.

The classification and coding systems of the ICD-9 have been retained in the ICD-NA for the sake of compatibility. Because several years have passed since the Ninth Revision of ICD, some of the terms may appear to neurologists to be obsolete. In such instances, these terms have been retained for the sake of compatibility, but there is an explanatory note, given in square brackets, discouraging their use.

Numbering system of the ICD-NA

Each main numerical heading in the ICD-NA is an ICD-9 code number at the 3-digit level. Titles for each of these numbers and for groups of

codes and main sections remain exactly the same as those given in the ICD-9.

However, the whole of the ICD-NA is based on 5- and 6-digit code numbers related to the ICD 3- and 4-digit codes in the following way:

The first 3 or 4 digits of any ICD-NA code number are those of the ICD-9. However, most of the fifth digits and all the sixth digits are exclusive to the ICD-NA. The fifth and sixth digits allow for increased specificity within the broader 3- and 4-digit categories. In the creation of the ICD-NA, certain terms have been added to those available in ICD-9, but the terms in ICD-9 and their codes were not modified. At the 4-digit level, therefore, all codes in the ICD-NA match the corresponding codes in ICD-9. This numbering system enables the relationship between the ICD-NA category and the parent category in the ICD to be established from the code itself, and should facilitate comparisons between statistics compiled according to the ICD-NA and, say, national morbidity statistics compiled according to ICD-9.

There is a set of supplementary codes for the classification of external causes of injury and poisoning (E-codes), and a specific system for coding the morphology of neoplasms (M-codes). These codes are taken from those given in ICD-9. Only codes thought to be pertinent to neurology are included. The E- and M-codes are not included in the index of the ICD-NA.

Use of non-numerical symbols within codes

The Ninth Revision of the ICD, and thus the ICD-NA, contain an innovation in that there are two codes for certain diagnostic descriptions that contain elements of information both about a localized manifestation or complication and about a more generalized underlying disease process. One of the codes—marked with a dagger (†)—is positioned in the part of the classification in which the diagnostic description is located according to the ICD principle, that relating to the underlying disease; the other code—marked with an asterisk (*)—is positioned in the chapter of the classification relating to the organ system to which the manifestation or complication relates. Thus tuberculous meningitis has its dagger code (013.0†) in the chapter for infectious and parasitic diseases, and its asterisk code (320.4*) in the nervous system chapter.

The use of asterisk coding is entirely optional. It should never be employed in coding the underlying cause of death (only dagger coding should be used for this purpose), but may be used in morbidity coding and in multiple-condition coding in relation to either morbidity or mortality. Any published tabulations of frequencies based on asterisk coding,

whether according to the detailed list or one of the short lists, should be clearly annotated: "Based on ICD [or ICD-NA] asterisk coding".

Thus, through the use of the above symbols, provision is made for recording neurological manifestations of a general disease or condition. All asterisk (*) and dagger (†) code pairs are cross-referenced to each other.

"-" indicates a space within a code that does not contain any number. For example, the code for spontaneous intracerebral hemorrhage is 431.-1; the "-" indicates that the first position to the right of the decimal point has no digit in ICD-9.

X indicates a space within a code that is supposed to contain a number. The actual number to be substituted is dictated by the specific set of instructions pertaining to that code. For example, the code for thrombosis of a precerebral artery is 443.X4. The number that replaces the "X" designates the particular artery involved.

<div align="center">INDEX</div>

The index to this volume is an alphabetic list of all items in the classification of the ICD-NA, together with the corresponding ICD-NA rubric. Items are generally listed in the format of "noun, adjective". Thus, "tuberculous meningitis" would be given in the index as "meningitis, tuberculous". All syndromes, diseases, etc. are listed alphabetically under the corresponding eponym. For example, Guillain-Barré syndrome will be found in the index under "Guillain-Barré".

The reader is CAUTIONED AGAINST USING THE INDEX FOR PURPOSES OF CODING. The index is simply intended as a guide to the appropriate place in the classification to be consulted for obtaining the proper code. The classification often contains explanatory notes regarding the condition and special rules of inclusion and exclusion that must be considered in choosing the correct code. Finally, the reader is reminded that terms provided in the index are not exhaustive. It is not possible to give every synonym for each listed condition because of limitations of space. However, every attempt has been made to include the most frequently used terms.

"NEC" is an abbreviation for "not elsewhere classified" and is added after terms classified to residual or unspecified categories and to terms in themselves ill-defined, as a warning that specified forms of the condition are classified differently.

RECOMMENDED USE OF THE ICD-NA

(1) Until becoming familiar with the ICD-NA, the user should consult the index, main headings, and inclusion/exclusion terms before recording a diagnosis.

(2) It is usual to reserve 8 and 9 in the first or second decimal place for "other" and "unspecified" classifications. The classification "other" is used for conditions that are specified, but not otherwise classified. The classification "unspecified" is used where there has been an omission in diagnosis, or where it is not possible to be specific.

(3) If it is desired to code a case in which the diagnosis is uncertain, the appropriate category indicating the general nature or site of the lesion must be used, with an "unspecified" classification.

(4) In cases where a diagnosis has not been established, the coding used should indicate the nature, type, or location of the lesion as narrowly as possible.

(5) Provision is made for recording neurological manifestations of a general disease or condition. Such manifestations are indicated by an asterisk (*) code and have a corresponding dagger (†) code to indicate etiology. All asterisk and dagger code pairs are cross-referenced to each other.

(6) Synonyms are provided in parentheses where there is some controversy or in deference to usage, but the title outside parentheses is preferred. It is hoped that a concerted attempt will be made in the not-too-distant future to standardize nomenclature, thus obviating the need for such synonyms.

(7) In the coding of neoplasms, those desiring more histologic specificity should use the M-codes given on pages 194-213.

(8) Only terms relating in some way to the nervous system and its diseases are included in ICD-NA. If a required term is missing from ICD-NA, then the full version of ICD-9 should be used. Every effort has been made to ensure such instances will occur infrequently.

(9) Users who encounter problems or difficulties in the application of this classification are asked to communicate their comments using the form at the end of this volume, to Dr J. M. Orgogozo, Unité de Pathologie Vasculaire, Centre hospitalier universitaire de Bordeaux, 33076 Bordeaux Cedex, France, and to the Neurosciences Programme, Division of Mental Health, World Health Organization, 1211 Geneva 27, Switzerland.