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Review / Derleme

The 11th Edition of the International Classification of Diseases and Related Health Problems: The Global Impact on the Future of Neurology for the Next Generation

Uluslararası Hastalıklar ve İlgili Sağlık Sorunları Sınıflandırmasının 11. Versiyonu: Gelecek Nesil için Nörolojinin Geleceği Üzerindeki Küresel Etki

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Abstract

The new 11th edition of the International Classification of Diseases and Related Health Problems is now online and open to use. The preparation started in 2007, and work continued until its implementation in 2022. As the classification is now online and open to comment, it is subject to possible alteration. The Neurology Topic Advisory Group worked to update the classification for the nervous system diseases in Chapter 8, and many changes were adopted. Moving all cerebrovascular diseases to neurology is perhaps the most important achievement. Another notable change is creating new codes for conditions such as Prion diseases and various genetic conditions. The classification also acknowledges that the causes of dementia are neurological.

Keywords: Neurology, classification, stroke, Prion diseases

Öz

Uluslararası Hastalıklar ve İlgili Sağlık Sorunları Sınıflandırması'nın 11. versiyonu artık çevrimiçi ve kullanıma açık. Hazırlıkları 2007 yılında başladı ve 2022 yılında uygulamaya geçilinceye kadar çalışmalar devam etti. Sınıflandırma artık çevrimiçi ve yoruma açık olduğundan olası değişikliklere tabidir. Nöroloji Konu Danışma Grubu, Bölüm 8'deki sinir sistemi hastalıkları sınıflandırmasını güncellemek için çalıştı ve birçok değişiklik kabul edildi. Tüm beyin damar hastalıklarının nörolojiye taşınması belki de en önemli başarıdır. Bir diğer dikkate değer değişiklik ise Prion hastalıkları ve çeşitli genetik durumlar için yeni kodların oluşturulmasıdır. Sınıflandırma ayrıca demansın nedenlerinin nörolojik olduğunu da kabul etmektedir.

Anahtar Kelimeler: Nöroloji, sınıflandırma, inme, Prion hastalıkları

Introduction

For the vast majority of clinicians, academic neurologists, and academic neurosurgeons, the idea of getting involved with classifying the diseases of the nervous system may seem tedious. Placing all nervous system diseases in logical categories and making the application of such a classification easy to use is daunting. It is also an evolving task, given that opinions on conditions change over time, and any classification must have a mechanism to allow evolution and change to happen in real time. Indeed, the task of moving from the rigid typed volumes of the 10th edition of the International Classification of Diseases and Related Health Problems (ICD-10) to the interactive 11th edition of the ICD (ICD-11) is an enormous one (1).

The international classification of diseases is the backbone of recognizing various illnesses and disorders. The classification system is a primary function of the World Health Organization (WHO), which started long before its establishment in 1948. The initial reporting was on causes of death that evolved into reporting on mortality and morbidity. The revision aimed to produce a modern classification to replace the ICD-10 that was established in the 1980s. Our knowledge of genetics, microbiology, immunology, imaging, therapeutics, and management has significantly changed our current medical practice (2).

In 1955, cerebrovascular diseases were reclassified as circulatory system diseases in the 7th edition of the ICD (ICD-7). The WHO's idea then was that a stroke is a condition affecting

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the blood vessels. This decision to reclassify cerebrovascular diseases to circulatory system diseases seemed contrary to the pathophysiology and symptoms leading to mortality and morbidity, which are those of brain dysfunction. Moreover, the decision deviated from the principle of ischemia in other organs (such as the intestines, kidneys, and the eye), which were listed under their respective organs in the ICD-7. Furthermore, various manifestations of cerebrovascular diseases were illogically placed in different parts of the ICD-10. Perhaps the most glaring example is that of transient ischemic attack put in a chapter on episodic symptoms!

Over the past six decades, this decision has skewed statistics because the 15 million people who had a stroke each year were included under the rubric of circulatory diseases. Stroke became a Cinderella disease, attracting little attention since it was thought that immediate treatment was not available. Treatment of stroke has been revolutionized over the past 20 years since the advent of stroke units, thrombolysis, and thrombectomy. Hyper acute stroke units save lives (3).

Stroke has been misplaced in the ICD since 1955, but it is now classed as a neurological disease in the new ICD-11. The reclassification required a bureaucratic struggle between clinicians and the WHO, but it should bring great benefits.

The WHO formally launched the process of revising the ICD-10 in 2007. A vast amount of work at the WHO headquarters and worldwide went into designing the development process. The program of work has been guided by regular meetings of representatives of the WHO Collaborating Centers for the Family of International Classifications, non-governmental organizations, and some other non-state actors, including the World Federation of Neurology, as well as the ICD-11 Revision Steering Group that supported the WHO through several special meetings, providing input on policy and content (4).

Extensive preparations were devoted to a review of the suitability of the structure of the ICD, which was a statistical classification of diseases and other health problems, to serve a wide variety of needs, including mortality and morbidity statistics, reimbursement, measuring the quality of care, improving patients' safety, monitoring primary care, and clinical recording. Many ICD Revision Topic Advisory Groups (TAGs), including ours, and the WHO departments for different chapters undertook technical work with crosscutting ICD revisions. The TAGs also examined information modeling, mortality, morbidity, quality, and safety.

The ICD-11 is designed to meet diverse users' needs and information technology demands. One important innovation is the use of electronic tools and platforms to support coding, translation, and testing. Regular revisions of the ICD are necessary to accommodate advances in medical knowledge.

The product of this ongoing revision is suitable for a digital environment and includes electronic tools for coding, browsing, translation, review, and mapping. The revised ICD has been designed to become interoperable with related classifications and terminologies. In addition, new approaches, such as tools for coding in low-resource environments, will be better integrated into the ICD.

Changing the placement of a disorder in the ICD is in the hands of statisticians and coders and involves changing computer systems and practices worldwide. The change also entails major financial commitments by users to accommodate it.

Our TAG produced all the necessary scientific reasoning as to why cerebrovascular diseases should be moved to Chapter 8 on nervous system diseases, with approval obtained from the Cardiology TAG. Following three years of discussions, the move was eventually approved by the WHO Division of Informatics and Statistics. However, the matter arose again in 2016 when the ICD-11 was being reviewed. The statisticians reverted to the stance that stroke would stay under the diseases of the vascular system chapter as it had been since the ICD-7. The decisions were made available on the ICD web platform in August 2016.

The reversal was not a result of any scientific objections but was made purely on statistical and coding grounds. Subsequently, papers in which the Neurology TAG and the WHO Statistics Department each stated their points of view were published in the Lancet side by side (5,6), leading to a stalemate that needed to be resolved. Further data on the etiopathology of stroke were provided, and the discussion culminated in a face-to-face meeting in Geneva between the Neurology TAG representatives and the then WHO Director of Information, Evidence, and Research, several advisers on statistics, and a representative of the Division of Mental Health and Substance Use. After a whole day of discussions, we were informed that a decision would be made and announced in due course, making it clear that the TAG's opinions are indeed only advisory. One must underline that the ICD is owned by the statisticians and not the Division of Mental Health and Substance Use, which was supportive, and without whose support the Division of Informatics and Statistics would not change its stance.

Three months later, we were informed that the ICD-11 team had agreed to move cerebrovascular diseases to Chapter 8 on nervous system diseases. This decision was momentous: after six decades of inaccurate statistics, brain diseases are finally placed correctly, and their devastating consequences shall be appropriately attributed. This change should bring about vast improvements in acute stroke care, as the statistics will show its devastating effects on the brain. The application of modern acute treatment within hours of ischemic stroke needs to be spread worldwide: urgent availability of imaging, thrombolysis, and thrombectomy are now a necessity and should be advocated to all governments as a priority. The devastating effects on brain function and the major disability produced by stroke can now be looked at as a top priority for healthcare funders. This decision was most encouraging because it exemplifies the willingness of our statistician colleagues to listen to sound scientific reasoning and interventions by health ministries and civil society and to act appropriately. For that, we are grateful. After 62 years in exile, we can say now that stroke is a brain disease (7).

The lesson from this process is that the clinicians who deal with the WHO need patience and tenacity. Epidemiologists run the WHO, and what may be obvious to a clinician may not be to the WHO.

Political appointees fill many decision-making positions to ensure representation from across the world, and technical advisers tend to move on fairly regularly, leading to a lack of continuity. This structure can lead to exasperation and abandonment of crucial issues.

Of course, producing an all-encompassing classification of diseases such as the ICD is a mammoth task, and we must be grateful to the Division of Informatics and Statistics at the WHO for their work, which has taken over a decade. Nevertheless,

the WHO has massive stature across the world. Its powers and reach are much greater than those of any highly-sighted research institution, both in the developed world and, even more so, in the developing world. The primary purpose of the WHO has been disease prevention, especially infections, but disease management is now being included in the organization's aims. To enter this realm successfully, the WHO needs a major change of culture and methods of operation. The advocacy of universal health coverage by the WHO Director–General, Tedros Adhanom Ghebreyesus, is laudable and crucial but surely requires closer involvement with best medical practice to facilitate the use of the highest level of medical technology globally. The huge advances in stroke management and the potential to save millions of people from death and disability should be a front-runner. One has to add that what is easily applicable to medical management in a developed setting is not applicable at all universally.

The Neurology TAG worked tirelessly to produce a modern classification so that health ministries can report to the WHO with data that are as accurate as possible and help healthcare funders justify funding for the care of patients with neurological conditions, particularly stroke. The Neurosciences TAG enlisted the help of many experts and organizations, and for the work on cerebrovascular diseases, we collaborated closely with the World Stroke Organization.

The 11th revision of the ICD benefited from remarkable commitments and contributions from clinical experts, statisticians, classification experts, and other users. The initial phase was driven by 30 committees and working groups, including 21 TAGs with clinical expertise in all key areas, including neurology. Over 7,000 revision proposals were received through an internet platform. The ICD-11 is unique as it builds its purpose-specific classifications from a foundation component. This foundation component is a database that includes 47,000 entities characterized by 13 properties, such as body system, causation, functional consequences, and manifestations. The foundation not only forms the basis for the conventional representation of the ICD, a tabular list of now 26 chapters, but it also provides the basis for specialty classifications, in which the categories can be arranged according to the requirements of the respective field (8).

For nervous system diseases, there are many highlights in the ICD-11, which makes it stand out (9); perhaps the most salient is cerebrovascular diseases, as discussed. With the support of several neurology and neurosurgery specialty organizations, the Neurology TAG worked for 10 years to produce the final version of the classification. The World Stroke Organization worked within the Neurology TAG to disentangle stroke and include it within Chapter 8 on nervous system diseases. This was only possible with the collaboration of cardiologists who felt similar to neurologists that stroke is a brain disease. It was important because the WHO Statistics Division was reluctant to change. This is understandable because it meant changing coding systems worldwide, which entails major changes in statistics departments and resetting computer programs. This was accomplished, and brain diseases will receive their rightful recognition as the second cause of death and the first cause of disability (10).

As stated above, stroke is the second most common cause of death worldwide and the top contributor to disability-adjusted life years. These statistics should make stroke care a top priority for healthcare providers (9).

However, neurological disorders are not high on the radar of the WHO because their burden has not been recognized as a major cause of death and disability. The problem has been the construction and application of the ICD, in which stroke had not been classified as a neurological disease. For the newly released ICD-11, however, we fought for stroke to be included in the neurology chapter, which will impact data reporting and benefit patient care and research (11). To understand this process of ICD revision, we have to consider how the WHO works. The organization's main drivers are member states and their health ministers. The six elected directors of the regional offices oversee the executive based in Geneva. National offices report to the WHO on the prevalence of all diseases in their countries according to the ICD coding system.

As stroke has been part of the vascular diseases section of the ICD, its effects have been reported with those of cardiac diseases and, as a result, have been lost in the mix. For example, in data published in the WHO European Health Report 2012, stroke was nowhere to be seen (8). Consequently, funding for neurological care was lost, and governments were unaware of the scale of the problem.

Another unintended consequence of the ICD has been the classification of non-communicable diseases (NCDs) for the WHO Global Action Plan for the Prevention and Control of Non-communicable Diseases 2013–2020.

The original idea was to target the adverse effects of tobacco, high salt intake, alcohol abuse, and a sedentary lifestyle. A Global Coordination Mechanism to tackle NCDs, created in 2014, was evolving to implement Article 3.4 of the United Nations Sustainable Development Goals, which aims to reduce mortality from NCDs by one-third by 2030 (12).

However, the four broad categories of NCDs chosen by the WHO were cancer, diabetes, cardiovascular diseases, and respiratory diseases; neither stroke nor any of the neurodegenerative diseases were initially included.

Of perhaps equal importance in explaining the difficulties of this process is that the structure of the WHO favors mental health: neurology still falls under the Division of Mental Health and Substance Use. The heads of the division have always been psychiatrists or psychologists with a public health background. In the past 15 years, only one medical officer has been appointed as the neurology focal point within the department. Only in the last three years has a small Brain Health Unit been established under the Department of Mental Health and Substance Use. Indeed, one may question if the major division should be brain health, given that mental health is a function of the brain! We are, however, where we are.

Now that the cerebrovascular disease placement has been rectified in the ICD-11 with stroke classified as a disease of the nervous system, it allows recognition by governments and funders. Moreover, all 5,000 diagnostic labels have short definitions, which helps to identify all conditions better. For example, the definition of cerebral ischemic stroke "8B11" is "*Acute focal neurological dysfunction caused by focal infarction at single or multiple sites of the brain. Evidence of acute infarction may come either from a) symptom duration lasting more than 24 hours or b) neuroimaging or other techniques in the clinically relevant area of the brain. The term does not include infarction of the retina".*

All titles and definitions in the ICD-11 are open to public comments, and a specialist Medical and Scientific Advisory

Committee responds to comments. It remains for all those involved in the production of the nervous system chapter to watch and see the implementation for all conditions, and this will form the basis of recognition, education, and future discussion. Furthermore, the ICD-11 is a digital platform that is ontology-based, and this will enhance discussion and constant review.

Work on the review of the ICD-10, looking at our section, started in 2009 and has been checked in detail.

Other conditions not identified in the ICD-10 were Prion diseases, labeled as "slow virus" infections. There were, however, some major issues that required compromise, such as the placement of dementia and the etiology of cognitive disorders and, similarly, functional/dissociative disorders (13). Moreover, for the first time, the ICD-11 neurology chapter has short definitions of all entities (14).

Highlights of updates in other sections of the ICD-11 include the possibility to report antimicrobial resistance, an updated classification of HIV, improved coding of diabetes and allergies, and the ability to describe patient safety events.

Over time, country uses of the ICD have moved beyond tracking mortality and now include morbidity statistics, health financing, research, and clinical care (15).

Change in the WHO happens slowly for obvious reasons. The ICD-10 was released nearly 30 years ago. The time has come to change and move on. The WHO TAG on Neurosciences was formed in 2009, and work started immediately. The whole of neurosciences were reviewed thoroughly, and a new classification was produced. The nervous system disease section is on the ICD-11 platform and is open for inspection and comment. Major advances in genetics, immunology, imaging, and therapeutics have changed the landscape beyond recognition. The ICD-11 also has allowed a most crucial readjustment related to stroke in all its guises (https://icd.who.int/browse11/l-m/en).

The WHO executive board approved the ICD-11 in February 2019, and the World Health Assembly subsequently approved it in May 2019. Implementation was then started in January 2022. All involved should see the fruits of their input in years to come.

Ethics

Peer-review: Internally peer-reviewed.

Conflict of Interest: Raad Shakir was chair of the WHO Neurosciences Topic Advisory Group ICD-11 2009–2019.

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