

The WHO ICD-11 Classification of Dermatological Disorders: a new comprehensive online skin disease taxonomy designed by and for dermatologists

Journal:	British Journal of Dermatology
Manuscript ID	BJD-2021-1354.R1
Wiley - Manuscript type:	Perspectives
Date Submitted by the Author:	13-Jul-2021
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JMA 10.9.21

Perspectives 21/1354

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DEAR EDITOR, In May 2019, the World Health Assembly officially adopted the 11th Revision of the *International Classification of Diseases* (ICD-11). When the current 10th Revision (ICD-10) was released some three decades ago, the world was at the beginning of the modern information technology era. The World Health Organization (WHO) had long recognized that the 'one-size-fits-all' nature of the ICD-10 and its inability to adapt to change seriously hampered its usefulness in healthcare research and management. The initial designs for ICD-11 were formulated by the WHO in 2007. Since then, many individuals from around the world have participated in its development. The ICD-11 is the first revision of the ICD that has had significant input from dermatologists. The International League of Dermatological Societies (ILDS) was invited by the WHO to participate in the revision from the outset, and dermatologists have been directly involved in redesigning it for 12 years, with an ongoing commitment to maintain and update it. From January 2022, the ICD-11 will replace the ICD-10 as the WHO's official statistical classification system; existing ICD-10 entities can be easily mapped within ICD-11.²

Sensibly, the direct replacement of the ICD-10 has been given a specific name appropriate to its functions, the *ICD-11 for Morbidity and Mortality Statistics* (ICD-11 MMS). The 'Diseases of the skin' chapter within the ICD-11 MMS has been fundamentally restructured and greatly expanded to provide a more logical framework within which to place skin disorders. Importantly, the sophisticated underlying polyhierarchical architecture of the ICD-11 means that diseases classified in one location can also be viewed elsewhere, where this makes sense. Thus, it is possible for cutaneous neoplasms to be parented both to 'Neoplasms' and to 'Diseases of the skin', although their primary statistical location remains within the 'Neoplasms' chapter.

However, the ICD-11 MMS is merely one of a range of ICD-11 classifications derived from an underlying comprehensive ICD foundation from which it is possible to create many customized classifications with either less (e.g. primary care) or more detail (e.g. specialist

domains such as dermatology) than is present in the MMS. This versatility is possible because each diagnostic concept is linked to a unique number [uniform resource identifier (URI)] by which it can be located in any ICD-11 classification into which it is incorporated. This has enabled the representation of skin diseases to be radically expanded within a newly designed dermatology-focused classification, the 'ICD-11 Classification of Dermatological Disorders' (ICDD).³ For the first time, rare or novel skin diseases can be represented individually in ICD within a structure containing more than 2000 dermatological entities, which is fully linked to the ICD-11 MMS (Figure 1).

The electronic platform on which all ICD-11 classifications are placed offers considerable advantages over what is provided for ICD-10.2 The key changes and refinements contained in ICD-11 of most relevance to dermatology include: (i) the facility to link ICD-11 directly to patient record systems, thus simplifying accurate diagnosis recording and enabling sophisticated data analysis; (ii) the production of a global dermatology-specific detailed classification of skin diseases (the ICDD) fully integrated into the ICD-11 electronic platform; (iii) a rebalancing of the ICD, with greater prominence given to morbidity, helping to highlight the impact of skin disease on health and well-being; (iv) an enhanced ability to code with precision and breadth, increasing the representation and visibility of rare skin diseases currently 'lost' within classifications that aggregate rather than differentiate; (v) the facility to refine diagnoses by linkage to metadata from the new ICD-11 Extensions database, which provides supplementary detail in a broad range of categories, including severity, occupational relevance, infectious agents, histopathology, substances (medicinal and nonmedicinal) and anatomical site; (vi) a completely new surface topography classification enabling precise recording of surface location; (vii) an allergen classification to help monitor global trends in allergy and the development of strategies to counter them;⁵ and (viii) the use of the ICDD by the ILDS to assemble a carefully curated reference gallery of images representing global skin disease in different skin types.

The ICD-11 MMS has already been field tested by dermatologists from across the world, confirming that an appropriate diagnostic term can be found for the overwhelming majority of patients presenting with skin disease. Further external review of the ICDD has been commissioned. While major structural changes to the ICD-11 will be limited and infrequent, it is vital that the international dermatology community continues to be involved in refining and enhancing the overall value and relevance of the classifications. A simple login is all that is required to submit proposals for change.⁶ All current versions of ICD-11 are available online at https://icd.who.int/en.

Acknowledgments

The authors gratefully acknowledge the significant debt of gratitude owed to the members of the Dermatology Topic Advisory Group and, in particular, to Professor Michael Weichenthal, Co-Chair of the project, whose input was crucial in creating and overseeing the classification and administrative frameworks for developing both *International Classification of Diseases*, 11th revision, for Mortality and Morbidity Statistics (ICD-11 MMS) 'Diseases of the skin' chapter and the ICD-11 Classification of Dermatological Disorders (ICDD).

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Conflicts of interest: the authors declare they have no conflicts of interest.

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Figure 1 Comparison of the representation of lichenoid dermatoses within the *International Classification of Diseases, 11th revision, for Mortality and Morbidity Statistics* (ICD-11 MMS) 4 (above) and the *ICD-11 Classification of Dermatological Diseases* (ICDD) 5 (right); the ICDD classification includes additional unique disorders that are part of the lichenoid dermatoses spectrum (i.e. lichen nitidus, lichen striatus, keratosis lichenoides chronica, druginduced oral lichenoid reaction and 'other' specified lichenoid dermatoses). Note that lichenoid drug eruption and drug-induced oral lichenoid reaction appear greyed out because the primary statistical location for these entities is 'Drug eruptions'.



- Papulosquamous dermatoses
 - ▶ EA90 Psoriasis
 - EA91 Lichen planus
 - ▼ EA92 Lichenoid dermatoses

EH62 Lichenoid drug eruption

EA93 Pityriasis lichenoides

EA94 Pityriasis rubra pilaris

EA95 Small plaque parapsoriasis

- Papulosquamous dermatoses
 - Psoriasis (EA90)
 - Lichen planus (EA91)
 - Lichenoid dermatoses

Lichen nitidus (EW1K DER)

Lichen striatus (EW1L DER)

Keratosis lichenoides chronica (EW1M DER)

▼ Lichenoid drug eruption (EH62)

Drug-induced oral lichenoid reaction (EW18 DER)

Other specified lichenoid dermatoses (EW1Z DER)

- Pityriasis lichenoides (EA93)
- Pityriasis rubra pilaris (EA94)

Small plaque parapsoriasis (EA95)