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Review

Developing classification criteria for discoid lupus erythematosus: an update from the World Congress of Dermatology 2015 meeting☆,☆☆

Scott A. Elman, AB [a], Filippa Nyberg, MD [b], Fukumi Furukawa, MD [c], Mark Goodfield, MD [d], Minoru Hasegawa, MD [e], Branka Marinovic, MD [f], Jacek Szepietowski, MD [g], Jan Dutz, MD, FRCPC [h], Victoria P. Werth, MD [i], Joseph F. Merola, MD, MMSc [a,j,*]

[a] Harvard Medical School, Boston, MA
[b] Institution for Clinical Sciences, Karolinska Institutet at Danderyd Hospital, Stockholm, Sweden
[c] Department of Dermatology, Wakayama Medical University, Wakayama, Japan
[d] Department of Dermatology, Leeds General Infirmary, Leeds, UK
[e] Department of Dermatology, Faculty of Medical Sciences, School of Medicine, University of Fukui, Fukui, Japan
[f] University Department of Dermatology and Venereology, Zagreb University Hospital Center and School of Medicine, Zagreb, Croatia
[g] Department of Dermatology, Venereology and Allergology, University of Medicine, Wroclaw, Poland
[h] Department of Dermatology and Skin Science, University of British Columbia, Vancouver, BC, Canada
[i] Philadelphia Department of Veterans Affairs Medical Center and Department of Dermatology, University of Pennsylvania, Philadelphia, PA
[j] Division of Rheumatology, Departments of Dermatology and Medicine, Brigham and Women's Hospital, Boston, MA

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ABSTRACT

Currently, no standardized classification criteria exist for cutaneous lupus erythematosus. With increased interest in studying cutaneous lupus erythematosus, specifically discoid lupus erythematosus, it is our aim to apply previously adopted methods from rheumatology to dermatologic diseases to develop feasible, validated, and standardized classification criteria useful in both academic and community practice. Here we report the progress to date to define discoid lupus erythematosus using clinical, histopathologic, and serologic features by means of a Delphi method—using a series of iterative questionnaires sent to expert stakeholders. We present specific updates from the World Congress of Dermatology 2015 meeting, at which a nominal group of expert stakeholders met to discuss the results of round 1 of the Delphi process to further clarify and harmonize specific classification items for inclusion into round 2.

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The need for clinicians who specialize in connective tissue diseases—adult and pediatric dermatologists, rheumatologists, and dermatopathologists—to create and agree upon a classification system for cutaneous lupus erythematosus (CLE), specifically discoid lupus erythematosus (DLE), was a topic of continued discussion at the World Congress of Dermatology (WCD), which was held June 2015 in Vancouver, Canada. At this meeting, a nominal group of stakeholders reviewed data from round 1 of the Delphi consensus process for designing a set of classification criteria for DLE; the classification criteria item list was reviewed for harmonization, distillation, and clarification before proceeding with round 2 of the process. Here we review the need for uniform classification criteria for CLE, why DLE was chosen as the first disease for classification criteria design, the results of the WCD meeting, and next steps in the process.

There is consensus—outlined previously—that CLE remains an ill-defined set of disorders that are often grouped together based on common features, and at present, inadequate definitions of CLE impede communication between physicians and in physician–patient interactions (Merola et al., 2015). Furthermore, a specific classification system that allows for identification of patients with CLE to

Abbreviations: ARS, Audience response system; CLE, Cutaneous lupus erythematosus; DLE, Discoid lupus erythematosus; ICCLE, International Conference on Cutaneous Lupus Erythematosus; SCLE, Subacute cutaneous lupus erythematosus; SLE, Systemic lupus erythematosus; WCD, World Congress of Dermatology.

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* Corresponding Author.
E-mail address: jfmerola@bwh.harvard.edu (J.F. Merola).

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facilitate further studies of these diseases does not exist. For these reasons, great heterogeneity exists in studies of CLE subsets, in particular among studies of discoid lupus cohorts, and underscores the need for classification criteria.

Classification criteria, the standardized definitions that are primarily intended to enable clinical studies to have uniform cohorts for research, require high specificity at the expense of some sensitivity. Classification criteria can be applied to different geographical regions, races, and ethnicities and do not require a gold standard test. Diagnostic criteria, however, reflect a broader and more variable set of features of a given disease; this is less ideal for identification of a uniform set of a study population.

The need for a more precise classification system for CLE was first formally discussed at the 3rd International Conference on Cutaneous Lupus Erythematosus (ICCLE), held May 2013 in Edinburgh, Scotland. The ultimate goal of this meeting was to agree on uniform definitions and grouping schemes and the understanding of the complex relationship between cutaneous and systemic disease. It was agreed that the Delphi technique—a method of consensus building using a series of iterative questionnaires to collect data from a panel of selected experts/stakeholders—would be utilized. The point of the Delphi technique is to achieve convergence of opinion on an issue. Briefly, experts anonymously answer questionnaires in two or more rounds; after each round, a facilitator provides an anonymous summary of the experts' thoughts from the previous round. Experts are encouraged to revise their earlier answers in light of the replies of other members. During this process, the range of answers decreases as the group converges toward one uniform answer. The benefits are subject anonymity (which reduces the effects of dominant individuals) and the inclusion of a geographically inclusive cohort due to the electronic nature of the survey. This process has been adopted previously by rheumatologists in creating outcome measures in rheumatoid arthritis and in developing feasible, valid, and standardized classification criteria for systemic sclerosis (Bartlett et al., 2012; Coulter et al., 2013).

A pre–Delphi questionnaire was sent to 81 stakeholders, including adult and pediatric dermatologists, rheumatologists, and dermatopathologists. The goal of the questionnaire was to query the participants regarding the need for uniform definitions and grouping schema for systemic lupus erythematosus and CLE and (1) to demonstrate consensus for a new definition of CLE, (2) to better clarify/define the relationship between CLE and systemic lupus erythematosus, and (3) to reevaluate current disparate grouping schema (Merola et al., 2015).

To begin the Delphi process to devise classification schema for CLE, it was decided that the first focus would be on DLE, with the idea of expanding the methodology to other subsets of CLE. DLE was specifically selected because there is increasing interest in understanding DLE's burden of disease, disease prevalence, and treatment of recalcitrant disease. For the aims of studying DLE epidemiology and treatment outcomes, it is important to better understand the description of DLE for most patients and to distinguish it from disease mimickers, such as (1) other cutaneous connective tissue disorders (e.g., dermatomyositis, subacute cutaneous lupus) and (2) other inflammatory dermatoses (e.g., tinea capitis, alopecia aerata, rosacea).

Based on discussions and breakout groups during the ICCLE meeting in 2013, round 1 of the Delphi questionnaire, the objective of which was to identify items for further evaluation as classification criteria for DLE, was created and distributed to 85 preidentified international experts in the field, including adult and pediatric rheumatologists, dermatologists, and dermatopathologists. The Delphi questionnaire comprised 48 items within the subcategories of morphology, histopathology, laboratory/serology, and symptomatology; all participants were asked to evaluate the appropriateness of the relevance of each characteristic to the diagnosis of DLE on a numerical scale from 0 to 100.

Sixty-one (72%) participants from Europe (31.0%), North America (60.3%), South America (1.7%), and Asia (6.9%) completed the round 1 questionnaire. Data interpretation was performed using prespecified cutoffs, using the median of the appropriateness ranking from 0 to 100, which effectively represents the percent consensus. It was deemed that consensus exists for items with medians ≥70 and ≤30; specifically, items with ≥70 were advanced to round 2, and those with ≤30 were removed from round 2 consideration. Those items with >30 but <70 were interpreted as having no consensus and technically did not meet criteria for round 2 advancement. However, these items were discussed among the expert panel at the WCD to ensure that clarification was not needed in the definition before removing items from round 2.

The initial results were presented for discussion at the WCD 2015 meeting in Vancouver. A subset of 30 participants were present, with the overall goal of determining which items that would have been eliminated based on statistical criteria alone should remain on the item list while minimizing the total number of items. The participants voted on item inclusion, provided justification for their choices, and were asked to provide feedback on item wording and to suggest new items for the item list. The responses from this meeting were recorded using an audience response system; although these responses do not change the questionnaire items directly, the information gathered will be used to inform the participants in the text of the round 2 questionnaire and allow the items to be voted on by a full group of stakeholders.

Ultimately, it is the goal of the Delphi process to apply previously adopted methods from rheumatology and other medical specialties to dermatologic diseases in order to develop feasible, validated, and standardized classification criteria useful in both academic and community practice. The classification criteria will be supported by further definitions and training modules to help nondermatologists and dermatologists alike identify disease characteristics specific for DLE.

With this goal in mind, the next steps of this process include (1) developing a final item list from the Delphi results to classify DLE and (2) validating this item list using strategies similar to those used in rheumatology to classify systemic sclerosis. Similar strategies should be embraced for more robust classification of other cutaneous diseases.

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