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Accessibility
Review

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

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A B S T R A C T

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...
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 individu-
als) 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 standard-
ized 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 dermat-
opathologists. 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 erythema-
osus, and (3) to reevaluate current disparate grouping schema
(Merola et al., 2015).
To begin the Delphi process to devise classification schema for CLE,
we decided that the first focus would be on CLE, with the idea of
expanding the methodology to other subsets of CLE. DLE was
specifically selected because there is increasing interest in under-
standing 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 areata, 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 Del-
phi 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
decided that consensus exists for items with medians ≥70 and ≤80;
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 nonrheumatologists
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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