Development of classification criteria for discoid lupus erythematosus: Results of a Delphi exercise

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Background: No classification criteria currently exist for discoid lupus erythematosus (DLE), which has led to problematic heterogeneity in both observational and interventional research efforts.

Objectives: We sought to develop DLE classification criteria based on consensus of international expert opinion of relevant stakeholders in the field.

Methods: Using a Delphi consensus process and nominal group techniques, potential items for classification criteria were generated. Experts ranked items in terms of their appropriateness and ability to discriminate DLE from other diagnoses, and items were subsequently eliminated using consensus exercises.

Results: A final list of 12 clinical and histopathologic items was generated for potential inclusion into a set of DLE classification criteria through a formal ongoing validation process.

Limitations: The participants are predominantly composed of DLE experts in North America and Europe.

Conclusion: This work represents a key step toward the development of formal DLE classification criteria. (J Am Acad Dermatol 2017;77:261-7.)

Key words: classification criteria; connective tissue diseases; cutaneous lupus erythematosus; Delphi method; discoid lupus erythematosus; lupus.

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There is consensus—outlined previously—that cutaneous lupus erythematosus (CLE) remains an ill-defined set of disorders that are often grouped together based on common features.[1] Currently, inadequate definitions of CLE impede communication between physicians and in physician-patient interactions. These sentiments were expressed at the Third International Meeting on CLE in 2013, during which an international group of lupus experts mutually agreed on the need for better definitions, grouping schema, and classification criteria for CLE variants.[1]

CAPSULE SUMMARY

- No classification criteria currently exist for discoid lupus erythematosus.
- Using an international Delphi consensus process, we identified a list of 12 potential items to serve as classification criteria for discoid lupus erythematosus.
- This work represents a key step toward the development of formal discoid lupus erythematosus classification criteria.

During the Third International Meeting on CLE, a decision was made to begin by developing classification criteria for discoid lupus erythematosus (DLE) for use in research endeavors. DLE, the most common type of chronic CLE, is a chronic dermatologic disease that can lead to scarring, hair loss, and dyspigmentation if not treated early or promptly. DLE was specifically chosen because it is considered one of the most common, readily recognizable, and potentially disfiguring forms of CLE. Furthermore, there is increasing interest in understanding DLE disease burden, disease prevalence, and the 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 (eg, dermatomyositis, subacute CLE); and (2) other inflammatory/infectious dermatoses (eg, psoriasis, tinea and other alopetic disorders, rosacea).

Importantly, there is currently no uniform definition of DLE on which to base a study population for observational and interventional trials; this has led to inconsistency among studies in the field. In addition, the current DLE grouping systems are heterogeneous, inconsistent, and none have been formally adopted by the "expert" community of investigators and clinicians committed to these disorders. Furthermore, variability in definitions impedes interpretation of findings from study to study and limits the ability to pool results and ultimately address questions of treatment efficacy.

Although a few authors proposed criteria for the diagnosis of DLE, none have proposed classification criteria.[2,3] It is important to make the distinction that classification criteria are the standardized definitions primarily intended to enable clinical studies to have uniform cohorts for research and require high specificity whereas diagnostic criteria reflect a more broad and variable set of features of a given disease.[4] The diagnostic criteria developed for DLE were derived from the authors' clinical expertise and were not validated. Furthermore, no comment has been made on the number of criteria that must be fulfilled to make a diagnosis of DLE.

Here we describe our efforts to identify a list of items for potential use as classification criteria and our next steps to validate these items. Our methodology has been largely adopted from the efforts of the American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) to develop classification criteria for systemic sclerosis, another connective tissue disease with a great deal of heterogeneity. Their process of identifying a list of items for potential classification criteria and deriving from them a criteria set have been described elsewhere.[5,6]

METHODS

To begin to devise classification criteria, a Delphi consensus approach was agreed on at the Third International Meeting on CLE in 2013. The Delphi technique is a method of consensus building using a series of questionnaires to a panel of selected experts and stakeholders; the iterative nature of this process, together with controlled anonymous feedback at each questionnaire stage, subject to anonymity and a predefined stop criterion, allow convergence toward a consensus.[7,8] Summary statistics of survey results are shared after each questionnaire, and experts are encouraged to revise their answers in light of responses from other members; during this process, the range of answers decreases as the group converges toward agreement of item utility. The benefits are subject anonymity and the inclusion of a geographically inclusive cohort. The study was approved by Partners/Brigham and Women's Institutional Review Board.

Design

This study had 2 phases: item generation followed by item reduction. An in-person approach was used for item generation, whereas internet-based Delphi consensus exercises and a face-to-face meeting using