AUTOIMMUNE CONNECTIVE TISSUE DISEASES

DEVELOPING CLASSIFICATION CRITERIA FOR CUTANEOUS DERMATOMYOSITIS

Js Concha [1] - J Merola [2] - D Fiorentino [3] - Jp Dutz [4] - M Goodfield [5] - F Nyberg [6] - B Volc-platzer [7] - M Fujimoto [8] - Cc Ang [9] - Vp Werth [1]

University Of Pennsylvania, Dermatology, Philadelphia, United States [1] - Brigham And Women's Hospital, Dermatology, Boston, United States [2] - Stanford University School Of Medicine, Dermatology, Redwood City, United States [3] - University Of British Columbia, Dermatology And Skin Science, Vancouver, Canada [4] - Leeds General Infirmary, Dermatology, Leeds, United Kingdom [5] - Karolinska University, Dermatology, Uppsala, Sweden [6] - Wiener Krankenanstaltenverbund, Dermatology, Vienna, Austria [7] - University Of Tsukuba, Dermatology, Tsukuba, Japan [8] - Changi General Hospital, Dermatology, Changi, Singapore [9]

Background: The new European League Against Rheumatism / American College of Rheumatology (EULAR / ACR) classification criteria for inflammatory myopathies are able to classify patients with cutaneous dermatomyositis (DM). However, approximately 25% of patients with skin-predominant DM do not meet 2 out of the 3 hallmark skin signs and fail to meet the criteria.

Objective: We aim to develop a set of skin-focused classification criteria that will distinguish cutaneous DM from mimickers and allow a more inclusive definition of skin-predominant disease.

Methods: An extensive literature review was done which guided international experts in generating items for the Delphi. Items were grouped into categories of distribution, morphology, symptoms, antibodies, histology, and contextual factors. Using REDCap™, participants rated these items in terms of appropriateness and distinguishing ability from mimickers. The relevance score ranged from 1 to 100, and the median of each item determined a rank-ordered list. A pre-specified median score cut-off was decided by the steering committee and the participants. There was a pre-Delphi and two rounds of actual Delphi. Resultant items underwent pilot validity testing using a single-center DM cohort.

Results: There were 50 dermatologists and rheumatologists from North America, South America, Europe, and Asia in this consensus exercise. After subjecting the items to a cutoff score of 70 during the first round, 37 out of the initial 54 items were retained and carried over to the next round. The cutoff was then raised to 80 during round two, and a list of 25 items was generated. Majority of the items displayed good face validity, being prevalent in a

cohort of diagnosed DM patients.

Conclusion: This project is a key step in the development of prospectively validated classification criteria that will create a more inclusive population of DM patients for clinical research on novel treatments for the disease.