Response to: 'Correspondence on 'EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups' by Irfan *et al*

We have with interest read the 'Correspondence on "EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups" by Irfan *et al*' with subtitle: improving the subtyping of dermatomyositis in the 2017 EULAR/ACR classification criteria for juvenile and adult idiopathic inflammatory myositis: value of non-classic cutaneous findings, to be published in *Annals of the Rheumatic Diseases*.¹ Here, the authors present performance of the EULAR/ACR classification criteria² in a cohort from India of 26 consecutive adult patients with idiopathic inflammatory myopathies (IIM). Patients with characteristic skin rashes (those included in the EULAR/ACR criteria as well as in the older Bohan and Peter criteria) were classified as having dermatomyositis (DM). The remainder of the patients were classified as polymyositis (PM) (n=12). All patients had documented muscle weakness. Of the 12 patients classified as PM, seven had non-classical skin rash such as generalised erythematous rash or malar rash and had been given the diagnosis DM by their physician. Moreover, five of these seven patients had myositis-associated autoantibodies that largely supported the clinical diagnosis of DM. The authors conclude that the sensitivities of Gottron's papules (60%–80%) and heliotrope rash (30%) are low. The authors suggest that ethnic or geographical differences in the prevalence of different skin rashes may explain this discrepancy in performance of the EULAR/ACR criteria in their cohort of Indian patients compared with the original cohort. They suggest that adding non-classic cutaneous findings for subclassification would substantially improve the sensitivity of the criteria for subtyping patients with DM across different populations.

We agree with the observed limitation of new EULAR/ACR classification criteria for DM. Other reports on the performance of the new criteria have described misclassification of DM with varying results. In one cohort with mainly Caucasian patients 3/151 patients who were clinically subclassified as DM did not have any of the EULAR/ACR criteria skin variables according to the EULAR/ACR criteria; these patients had typical shawl or V-sign.³ In a cohort from China, 7 of 40 patients with a clinical diagnosis of DM were classified as PM using the EULAR/ACR criteria.⁴ These patients had shawl sign, holster sign, erythroderma or other rashes frequently seen in DM patients. In a recent study of 194 adult Japanese patients diagnosed as DM by the physician, 14 were classified as PM by applying the EULAR/ACR criteria.⁵ In a cohort of 99 patients with well-characterised amyopathic DM (ADM) from a dermatology centre, 6% did not have any of the 'classic skin rashes' and 26% could not be classified as ADM/DM using the EULAR/ACR criteria. In this cohort, Gottron's sign was the most common skin rash (present in 90%) followed by malar rash (82%), periungual changes (77%), V-area of the neck (76%) and rest of the face rash (76%). Sixty-two percent had a heliotrope, similar to what was found in the international population used to derive the EULAR/ACR criteria.⁶

In the development of the EULAR/ACR classification criteria, 93 variables were tested including 14 skin variables. In addition to the three classical skin rashes, data were collected on shawl and V-sign rashes, periorbital oedema, linear extensor erythema, calcification, periungual erythema, mechanic's hands, photodistributed violaceous erythema, Raynaud's phenomenon, cuticular

Figure 1 Differences in the prevalence of skin manifestations between Caucasian and Asian dermatomyositis (DM) and amyopathic DM (ADM) patients based on data from the International Myositis Classification Criteria Project.² P values are derived from significance of difference between Caucasian DM patients and Asian DM patients, and between Caucasian ADM patients and Asian ADM patients, respectively. *p<0.05, **p<0.01, ***p<0.001, ****p<1E-5.

overgrowth and poikiloderma.² In the statistical analyses based on 976 IIM patients (DM, n=239; ADM, n=44; hypomyopathic DM, n=12; juvenile (DM) (JDM), n=248), and 624 non-IIM patients, only the three classical skin rashes were found to be significant in the classification of IIM from non-IIM patients, and were included in the final version of the criteria.

A difference in the presence of DM-associated skin variables in various ethnic populations is suggested by data from the classification criteria cohort.⁷ In the DM subgroup, seven skin manifestations were more prevalent among Caucasian compared with Asian patients, whereas shawl-sign was more common among Asian patients. In the subgroup ADM, Caucasian patients had a higher prevalence of seven skin manifestations compared with Asian patients. Still the classical skin rashes were among the most common in both Asian and Caucasian patients (figure 1). Among DM patients, 9.6% were negative for all three EULAR/ACR criteria skin rashes and among the ADM patients no patient was negative for all three.

As discussed in previous letters to *Annals of the Rheumatic Diseases*, the EULAR/ACR criteria have a limitation as most myositis-specific autoantibodies, except anti-Jo1 autoantibodies, were not available to be included in the criteria due to historical reasons.^{8–10} Thus, an update of the criteria is needed. To test the performance of the whole panel of myositis-specific autoantibodies, and potentially other variables, we welcome new collaborators with large cohorts of patients with IIM and comparator cases with validated results of autoantibody profiles, as well as thorough clinical data on muscle and extra-muscular variables, including skin rashes. An ongoing collaborative effort between dermatology and rheumatology to examine prospectively collected skin data in DM will further examine the skin variables.¹¹

Ingrid E Lundberg¹,¹ Frederick W Miller²,² Lisa G Rider,² Victoria P Werth^{3,4},^{3,4} Anna Tjärnlund,¹ Matteo Bottai⁵

¹Division of Rheumatology, Department of Medicine, Solna, Karolinska Institutet, and Karolinska University Hospital, Stockholm, Sweden

²Environmental Autoimmunity Group, Clinical Research Branch, National Institute of Environmental Health Sciences, National Institutes of Health, Bethesda, Maryland, USA

³Dermatology, University of Pennsylvania Perelman School of Medicine, Philadelphia, Pennsylvania, USA

⁴Medicine, Corporal Michael J Crescenz VA Medical Center, Philadelphia, Pennsylvania, USA

⁵Institute of Environmental Medicine, Karolinska Institutet, Stockholm, Sweden

Correspondence to Professor Ingrid E Lundberg, Rheumatology, Karolinska Institute, Stockholm SE-17176, Sweden; ingrid.lundberg@ki.se

Handling editor Josef S Smolen

Contributors All authors were involved in drafting the article or revising it critically for important intellectual content and approved the final version to be published. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Funding This research was supported in part by the Intramural Research Program of the NIH, National Institute of Environmental Health Sciences.

Competing interests None declared.

Patient and public involvement Patients and/or the public were not involved in the design, or conduct, or reporting, or dissemination plans of this research.

Patient consent for publication Not required.

Provenance and peer review Commissioned; internally peer reviewed.

© Author(s) (or their employer(s)) 2021. No commercial re-use. See rights and permissions. Published by BMJ.

To cite Lundberg IE, Miller FW, Rider LG, *et al.* *Ann Rheum Dis* Epub ahead of print: [please include Day Month Year]. doi:10.1136/annrheumdis-2020-219436

Received 24 November 2020

Accepted 25 November 2020

► <http://dx.doi.org/10.1136/annrheumdis-2020-219426>

Ann Rheum Dis 2021;0:1–2. doi:10.1136/annrheumdis-2020-219436

ORCID iDs

Ingrid E Lundberg <http://orcid.org/0000-0002-6068-9212>

Frederick W Miller <http://orcid.org/0000-0003-2831-9593>

Victoria P Werth <http://orcid.org/0000-0003-3030-5369>

REFERENCES

- 1 Irfan M, Liza R. Correspondence on "EULAR/ACR Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies and their Major Subgroups by "Ingrid E Lundberg, *et al.* *Ann Rheum Dis* 2020. doi:10.1136/annrheumdis-2020-219426
- 2 Lundberg IE, Tjærnlund A, Bottai M, *et al.* 2017 European League against Rheumatism/American College of rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. *Ann Rheum Dis* 2017;76:1955–64.
- 3 Barsotti S, Dastmalchi M, Notarnicola A, *et al.* Performance of the new EULAR/ACR classification criteria for idiopathic inflammatory myopathies (IIM) in a large monocentric IIM cohort. *Semin Arthritis Rheum* 2020;50:492–7.
- 4 Zhang X, Yang X, Ji L, *et al.* Validation of 2017 classification criteria for adult and juvenile idiopathic inflammatory myopathies proposed by EULAR/ACR in Chinese patients. *Int J Rheum Dis* 2019;22:1278–82.
- 5 Jinnin M, Ohta A, Ishihara S, *et al.* Research Team for Autoimmune Diseases, The Research Program for Intractable Disease of the Ministry of Health, Labor and Welfare. First external validation of sensitivity and specificity of the European League Against Rheumatism (EULAR)/American College of Rheumatology (ACR) classification criteria for idiopathic inflammatory myopathies with a Japanese cohort. *Ann Rheum Dis* 2020;79:387–92.
- 6 Patel B, Khan N, Werth VP. Applicability of EULAR/ACR classification criteria for dermatomyositis to amyopathic disease. *J Am Acad Dermatol* 2018;79:77–83.
- 7 Tjærnlund A, Rider LG, Miller FW, *et al.* Ethnic but not gender differences in disease manifestations in dermatomyositis patients. *Arthritis Rheum* 2011;63:S87.
- 8 Lundberg IE, Tjærnlund A. Response to: '2017 EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups: little emphasis on autoantibodies, why?' by Malaviya. *Ann Rheum Dis* 2018;77:e78.
- 9 Tjærnlund A, Rönnelid J, Bottai M, *et al.* Response to: 'Detection of myositis-specific antibodies' by Vulsteke *et al.* *Ann Rheum Dis* 2019;78:e8.
- 10 Tjærnlund A, Lundberg IE. Response to: 'Comment on: 'Idiopathic inflammatory myopathies and antisynthetase syndrome: contribution of antisynthetase antibodies to improve current classification criteria' by Greco *et al.* by Knitza *et al.* *Ann Rheum Dis* 2020;79:e86.
- 11 Concha JSS, Pena S, Gaffney RG, *et al.* Developing classification criteria for skin-predominant dermatomyositis: the Delphi process. *Br J Dermatol* 2020;182:410–7.