Response to: 'Performance of the 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies in clinical practice' by Hočevar *et al*

We read with interest the letter titled 'Performance of the 2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies in clinical practice' by Hočevar *et al* published in the *Annals of the Rheumatic Diseases*.¹ In the letter the authors report the sensitivity and specificity of the recently published '2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups'² observed in a retrospective cohort of 95 patients with idiopathic inflammatory myopathies (IIM) collected between 2010 and 2017 in a Slovenian rheumatology centre. As comparators, they used 72 subjects who had had a work-up for IIM, including muscle biopsy, during the same time period as the patients with IIM but without proof of IIM. The sensitivity and specificity were obtained for both the model with and that without muscle biopsy data. The sensitivity in the Slovenian rheumatology cohort was 74.7% without muscle biopsy data and 80.5% with muscle biopsy data. The corresponding figures reported in the original paper² were 87% and 93%, respectively. The specificity observed in the Slovenian rheumatology centre cohort was 80.6% without muscle biopsy data and 90.3% with muscle biopsy data. The corresponding figures reported in the original paper² were 82% and 88%, respectively. The positive and negative predictive values observed in the Slovenian cohort were also smaller than in the original paper²: positive predictive values of 84%–91% vs 90%–94%, and negative predictive values of 71%–79% vs 79%–85%.

The cases that were identified as the newly defined subgroup of IIM called immune-mediated necrotising myopathies in the Slovenian cohort were misclassified by the new criteria. This subgroup was very small (n=11) in the cohort used to derive the criteria. The subjects with a mild or no myopathy but with interstitial lung disease and myositis-specific autoantibodies (MSAs) other than anti-Jo-1 were also misclassified. This group was also small in the cohort that was used to derive the new criteria due to missing information on a majority of the new MSAs. It is not clear from the report by Hočevar *et al* how many cases had missing data and how this problem was handled. It is unclear to us how many patients were unclassifiable and how these were handled in the calculations. Unclassifiable patients are those whose predicted probability range spans across the cut-off value. Incorrect handling of these may help

explain the slightly smaller sensitivity and specificity observed in the Slovenian cohort as compared with the classification criteria cohort.

The report in the letter emphasises the need to validate the criteria in external cohorts including all aspects of the heterogeneous IIM population and also the need to revise the '2017 European League Against Rheumatism/American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups', including the newly identified subgroups of IIM like the immune-mediated necrotising myopathy and the patients with predominating extra-muscular manifestations such as interstitial lung disease associated with the newly identified MSAs using validated antibody assays. These opportunities for improvement have been emphasised in the original paper² and are on the research agenda.

We welcome this report and all others that may appear in the future that will contribute to the validation of the new criteria in external cohorts of patients.

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Handling editor Josef S Smolen

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Competing interests None declared.

Provenance and peer review Commissioned; internally peer reviewed.

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To cite Lundberg IE, Bottai M, Tjärnlund A. *Ann Rheum Dis* 2018;**77**:e91.

Received 20 December 2017

Accepted 23 December 2017

Published Online First 9 January 2018

► <http://dx.doi.org/10.1136/annrheumdis-2017-212774>

Ann Rheum Dis 2018;**77**:e91. doi:10.1136/annrheumdis-2017-212786

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