Response to: 'Do the 2019 EULAR/ACR SLE classification criteria close the door on certain groups of SLE patients?' by Chi *et al*

In their letter, Dr Chi and colleagues[1] express two concerns regarding the new European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria. On the one hand, they correctly remark that patients with uncommon, isolated organ manifestations may be more difficult to classify with any set of classification criteria. On the other hand, they caution against mistaking malignancies for systemic lupus erythematosus (SLE).

The first sensitivity-related issue is certainly correct. However, it is important to reiterate that the new EULAR/ACR criteria are classification criteria. Our manuscript explicitly states that they should not be used as diagnostic criteria and that diagnosis remains with the appropriately trained physician.[2 3] In order for a criteria set to remain feasible, that is, not to contain hundreds of items, uncommon isolated manifestations or laboratory tests cannot be accounted for. We had a focus on early symptoms,[4] where only fever qualified as sufficiently performing for classification in early disease to be added to the criteria list. Still, it is important to be aware of non-criteria organ manifestations to fully understand the complexity of SLE, and we thank our colleagues for bringing this up. As SLE is a highly heterogeneous disease, there are probably hundreds of rare SLE manifestations that are not included in the new criteria set, yet, the new criteria still have excellent sensitivity when compared with prior criteria sets.

For the second specificity-related issue, it is also important to distinguish classification from diagnosis. In addition, this gives us the opportunity to reiterate two important features of the new criteria[1 2] that should limit the risk of classifying patients with malignancies as patients with SLE. One, most of the malignancies that are mimicking SLE features are haematological. In this regard, going back to domains and counting the highest ranking item in each domain only is an advantage. This step, which followed an in-depth evaluation of associations between SLE items,[5] will prevent counting several changes in blood counts.

Even more important is the general attribution rule of the new criteria: items are only to be counted for SLE, if there is no more likely alternative explanation.[2 3] This approach requires both clinical understanding and diligence but should also safeguard against misclassifying malignancies.

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