Dr. Hoffmann-Vold replies

To the Editor:

We highly appreciate the interest[1,2] in our paper[3] and the reflected and sagacious views from Baron, *et al*[4]. As mentioned, it is still unclear whether mixed connective tissue disease (MCTD) is a distinct disease or might represent an overlap disease. Therefore, we were interested in reviewing the 16 patients with systemic sclerosis (SSc) of our cohort who did not fulfill the 2013 American College of Rheumatology (ACR)/European League Against Rheumatism (EULAR) criteria[5,6] and the 18 patients with MCTD who did fulfill the criteria in our original paper. Overall, we found that all patients with limited cutaneous SSc, none of the patients with sine scleroderma, and 3/6 patients with pre-scleroderma met the 2013 ACR/EULAR criteria at the last annual followup visit. When looking at the patients with MCTD, 4 patients were deceased. Of the remaining patients, 1 patient was diagnosed with SSc and 4 patients were clinically assessed as MCTD/SSc overlap and still met the 2013 ACR/EULAR criteria. Somewhat surprisingly, several patients did not fulfill the 2013 criteria at the followup visit, mainly because of different assessment of skin thickening of the fingers.

Fourteen of these 18 patients with MCTD were included in the HLA profile study of Norwegian patients with MCTD by Flåm, *et al*[7]. Of those, 7 had HLA alleles associated with MCTD and only 1 had HLA alleles associated with SSc.

Altogether, we believe that our study and the followup data clearly show that there is a need for further research on anti-RNP–positive patients, both in terms of genotypes and phenotypes.

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