EULAR Sjögren’s syndrome disease activity index: development of a consensus systemic disease activity index for primary Sjögren’s syndrome

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ABSTRACT

Objective To develop a disease activity index for patients with primary Sjögren’s syndrome (SS): the European League Against Rheumatism (EULAR) Sjögren’s syndrome disease activity index (ESSDAI).

Methods Thirty-nine SS experts participated in an international collaboration, promoted by EULAR, to develop the ESSDAI. Experts identified 12 organ-specific ‘domains’ contributing to disease activity. For each domain, features of disease activity were classified in three or four levels according to their severity. Data abstracted from 96 patients with systemic complications of primary SS were used to generate 702 realistic vignettes for which all possible systemic complications were represented. Using the 0–10 physician global assessment (PhGA) scale, each expert scored the disease activity of five patient profiles and 20 realistic vignettes. Multiple regression modelling, with PhGA used as the dependent variable, was used to estimate the weight of each domain.

Results All 12 domains were significantly associated with disease activity in the multivariate model, domain weights ranged from 1 to 6. The ESSDAI scores varied from 2 to 47 and were significantly correlated with PhGA for both real patient profiles and realistic vignettes ($r=0.61$ and $r=0.58$, respectively, $p<0.001$). Compared with 57 (59.4%) of the real patient profiles, 468 (66.7%) of the realistic vignettes were considered likely or very likely to be true.

Conclusion The ESSDAI is a clinical index designed to measure disease activity in patients with primary SS. Once validated, such a standardised evaluation of primary SS should facilitate clinical research and be helpful as an outcome measure in clinical trials.