Sjögren’s Syndrome Disease Damage Index and Disease Activity Index Scoring Systems for the Assessment of Disease Damage and Disease Activity in Sjögren’s Syndrome, Derived From an Analysis of a Cohort of Italian Patients

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Objective.
To develop valid instruments for the assessment of disease-related damage and disease activity in Sjögren’s syndrome (SS).

Methods. Data on 206 patients with primary SS were collected in 12 Italian centers. Each patient was scored by 1 investigator, on the basis of a global assessment of the degree of disease damage and disease activity. Patients judged to have active disease at the time of enrollment underwent a second evaluation after 3 months. Univariate and multivariate analyses were performed to select the clinical and serologic variables that were the best predictors of damage and of disease activity, and these variables were used to construct the Sjögren’s Syndrome Disease Damage Index (SSDDI) and the Sjögren’s Syndrome Disease Activity Index (SSDAI). The weight of each variable in the index was determined by the coefficients in multivariate regression models. Scores obtained using the SSDDI and the SSDAI were compared with scores initially given by the investigators. Finally, a receiver operating characteristic (ROC) curve was used to determine the cutoff value in the SSDAI with the highest level of accuracy in identifying patients with a significant level of disease activity.

Results. A multivariate model with 9 variables was the best predictor of investigator scores of damage. The scores obtained using the SSDDI were closely correlated with investigator ratings \( R = 0.760, P < 0.0001 \). A model composed of 11 variables was the best predictor of investigator scores of disease activity. The scores obtained using the SSDAI were strongly correlated with the investigator ratings both at the time of enrollment and 3 months after enrollment \( R = 0.872, P < 0.0001 \), and \( R = 0.817, P < 0.0001 \), respectively. The differences between scores given by investigators at study enrollment and after 3 months, a measure of variation of disease activity over time, were also closely correlated with the differences calculated using the SSDAI \( R = 0.683, P < 0.0001 \). The ROC curve analysis showed that patients with the highest level of disease activity could be identified on the basis of an SSDAI score of > 5.

Conclusion.
Our findings indicate that the SSDDI is an adequate instrument to objectively measure damage in patients with SS, and that the SSDAI is a valid tool to measure disease activity when used either as a single-state index or as a transition index.