Table 4. Sjögren's Syndrome Disease Activity Index*

Item	Definition	Score
Constitutional symptoms		
Fever	≥38°C, not due to infections	1
Fatigue	Sufficiently severe to affect normal activities	1
Change in fatigue	New appearance or worsening of fatigue	1
Change in salivary gland swelling	New appearance or increasing swelling of major salivary glands, not due to infection or stones	3
Articular symptoms (any of the following)		2
Arthritis	Inflammatory pain in ≥1 joint†	
Evolving arthralgias	New appearance or worsening of joint pain without signs of articular inflammation†	
Hematologic features		
Leukopenia/lymphopenia	<3,500 mm^3/<1,000 mm^3	1
Lymph node/spleen enlargement	Clinically palpable lymph node/spleen	2
Pleuropulmonary symptoms (any of the following)		4
Pleurisy	Confirmed by imaging, not due to infection	
Pneumonia (segmental or interstitial)	Ground-glass appearance on computed tomography scan, not due to infection	
Change in vasculitis	New appearance or worsening or recurrent flares of palpable purpura	3
Active renal involvement (any of the following)		2
New or worsening proteinuria	>0.5 gm/day	
Increasing serum creatinine level	Above the normal limits	
New or worsening nephritis	Glomerular or interstitial, histologically defined	
Peripheral neuropathy	Recent onset (<6 months), confirmed by nerve conduction studies	1

* The index was constructed using variables selected by means of a multivariate linear regression model. The score value assigned to each item was derived from the weight that the corresponding variable had in the model (β coefficient shown in Table 3). The definitions shown were provided in the glossary included with the clinical chart in which patient data were recorded.
† Excluding other causes of joint/muscle pain, such as osteoarthritis or fibromyalgia.

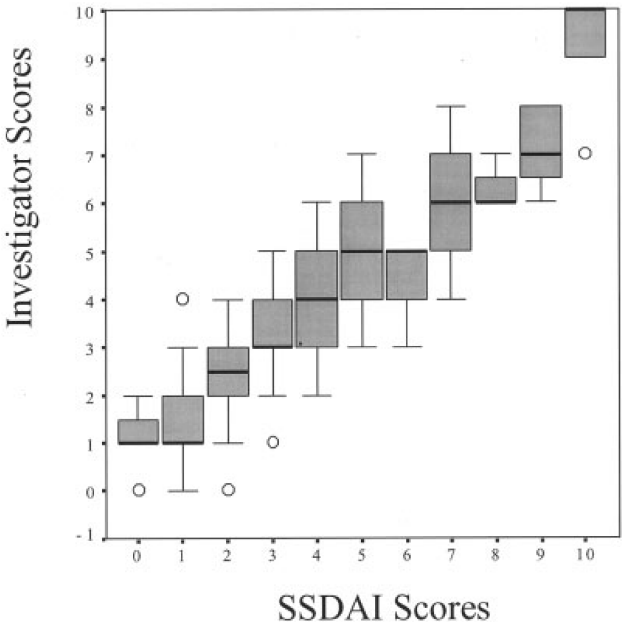

Figure 2. Correlation between the scores given by the investigators at the first observation time and those calculated using the Sjögren's Syndrome Disease Activity Index (SSDAI), in 206 patients with primary SS. The correlation was statistically significant (R = 0.872, $P < 0.0001$). Data are presented as box plots, where the boxes represent the 25th to 75th percentiles, the lines within the boxes represent the median, and whiskers represent the 10th and 90th percentiles. Circles indicate outliers.

were closely correlated (R = 0.683, $P < 0.0001$). The differences between the change in investigator scores from the first evaluation to the second evaluation and the change in SSDAI scores were also normally distributed ($W = 0.958$, $P < 0.001$). Values were outside the limit for 95% agreement (mean ± SD −0.24 ± 1.68) in only 6.6% of the patients.

Finally, the accuracy of the SSDAI in distinguishing patients initially classified by the investigators as having active or very active disease from those classified as having inactive or mildly/moderately active disease was assessed by ROC curve analysis. For this purpose, an SSDAI score of ≥5 had high sensitivity (86.5%) and specificity (87.6%).

DISCUSSION

In this study levels of disease damage and disease activity in patients with primary SS were evaluated. The clinical and laboratory variables that were the best predictors of disease damage or disease activity, as globally assessed by the investigators, were used to create 2 scoring systems, the SSDDI and the SSDAI. The construct validity of each instrument was confirmed by the correlation of index scores with the physician's global assessment of the respective disease state. Although the correlation between the scores derived using