2016 American College of Rheumatology/European League Against Rheumatism classification criteria for primary Sjögren's syndrome: A consensus and data-driven methodology involving three international patient cohorts.

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Abstract

OBJECTIVES:
To develop and validate an international set of classification criteria for primary Sjögren's syndrome (SS) using guidelines from the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR). These criteria were developed for use in individuals with signs and/or symptoms suggestive of SS.

METHODS:
We assigned preliminary importance weights to a consensus list of candidate criteria items, using multi-criteria decision analysis. We tested and adapted the resulting draft criteria using existing cohort data on primary SS cases and non-SS controls, with case/non-case status derived from expert clinical judgement. We then validated the performance of the classification criteria in a separate cohort of patients.

RESULTS:
The final classification criteria are based on the weighted sum of five items: anti-SSA/Ro antibody positivity and focal lymphocytic sialadenitis with a focus score of ≥1 foci/4 mm², each scoring 3; an abnormal Ocular Staining Score of ≥5 (or van Bijsterveld score of ≥4), a Schirmer's
test result of ≤5 mm/5 min and an unstimulated salivary flow rate of ≤0.1 mL/min, each scoring 1. Individuals with signs and/or symptoms suggestive of SS who have a total score of ≥4 for the above items meet the criteria for primary SS. Sensitivity and specificity against clinician-expert-derived case/non-case status in the final validation cohort were high, that is, 96% (95% CI 92% to 98%) and 95% (95% CI 92% to 97%), respectively.

**CONCLUSION:**
Using methodology consistent with other recent ACR/EULAR-approved classification criteria, we developed a single set of data-driven consensus classification criteria for primary SS, which performed well in validation analyses and are well suited as criteria for enrolment in clinical trials.