

6 Table. Classification Criteria for Sjogren's Syndrome: A Revised Version of the European Criteria Proposed by the American-European Consensus Group

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This is the standard current criteria used in clinical studies and in Europe. A separate criteria was proposed by Shiboski et al and was called the ACR criteria. The two criteria were about 90% similar but had important difference. A series of meetings have led to agreement for a new consensus criteria that will be published during 2016

I. Ocular symptoms: a positive response to at least one of the following questions:

1. Have you had daily, persistent, troublesome dry eyes for more than 3 months?
2. Do you have a recurrent sensation of sand or gravel in the eyes?
3. Do you use tear substitutes more than 3 times a day?

II. Oral symptoms: a positive response to at least one of the following questions:

1. Have you had a daily feeling of dry mouth for more than 3 months?
2. Have you had recurrently or persistently swollen salivary glands as an adult?
3. Do you frequently drink liquids to aid in swallowing dry food?

III. Ocular signs - that is, objective evidence of ocular involvement defined as a positive result for at least one of the following two tests:

1. Shirmer's test, performed without anaesthesia (  $\leq 5$  mm in 5 minutes )
2. Rose bengal score or other ocular dye score (  $\geq 4$  according to van Bijsterveld's scoring system )

III. Histopathology: In minor salivary glands (obtained through normal appearing mucosa ) focal lymphocytic sialoadenitis, evaluated by an expert histopathologist, with a focus score  $\geq 1$ , defined as a number of lymphocytic foci (which are adjacent to normal-appearing mucous acini and contain more than 50 lymphocytes) per 4 mm<sup>2</sup> of glandular tissue

IV. Salivary gland involvement: objective evidence of salivary gland involvement defined by a positive result for at least one of the following diagnostic tests:

1. Unstimulated whole salivary flow (  $\leq 1.5$  ml in 15 minutes )
2. Parotid sialography showing the presence of diffuse sialectasias (punctate, cavitory, or destructive pattern ), without evidence of obstruction in the major ducts.
3. Salivary scintigraphy showing delayed uptake, reduced concentration and/or delayed excretion of tracer

Autoantibodies: presence in the serum of the following autoantibodies:

1. antibodies to Ro(SSA) or La(SSB) antigens, or both

#### VI. Revised rules for classification

##### A. For primary SS

In patients without any potentially associated disease, primary SS may be defined as follows:

- a. The presence of any 4 of the 6 items is indicative of primary SS, as long as either item IV (Histopathology) or VI (Serology) is positive.
- b. The presence of any 3 of the 4 objective criteria items (that is, items I I I, I V, V, V I)
- c. The classification tree procedure represents a valid alternative method for classification, although it should be more properly used in clinical epidemiological survey.

##### B. For secondary SS

In patients with a potentially associated disease (for instance, another well defined connective tissue disease), the presence of item I or item I I plus any 2 from among items I I I, I V, and V may be considered as indicative of secondary SS.

#### III. Exclusive criteria:

Past head and neck radiation treatment

Hepatitis C infection

Acquired immunodeficiency disease (AIDS)

Pre-existing lymphoma

Sarcoidosis

Graft versus host disease

Use of anticholinergic drugs (since a time shorter than 4-fold life of the drug)

1. Vitali, C., S. Bombardieri, R. Jonsson, H. M. Moutsopoulos, E. L. Alexander, S. E. Carsons, T. E. Daniels, P. C. Fox, R. I. Fox, S. S. Kassan, S. R. Pillemer, N. Talal, and M. H. Weisman. 2002. Classification criteria for Sjogren's syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. *Ann Rheum Dis* 61:554-558.