



DIAGNOSTIC CRITERIA

SJÖGREN SYNDROME

INTRODUCTION:

Sjögren syndrome (SS) is a multisystem autoimmune disease characterized by hypo function of salivary and lacrimal glands and possible systemic multi-organ manifestations. It is primarily managed by rheumatologist, in collaboration with ophthalmologists and oral medicine/pathology specialists.

This criteria set has been approved by the American College of Rheumatology (ACR) Board of Directors and the European League Against Rheumatism (EULAR) Executive Committee. This signifies that the criteria set has been quantitatively validated using patient data, and it has undergone validation based on an independent data set.

- Sensitivity: 96%
- Specificity: 95%

REFERENCE VALUES:

- Not classified as primary SS: < 4
- Classified as primary SS: ≥ 4

Inclusion criteria

Positive response to a least one question

Have you had daily, persistent, troublesome dry eyes for more than 3 months

Do you have a recurrent sensation of sand or gravel in the eyes?

Do you use tear substitutes more than three times a day?

Have you had a daily feeling of dry mouth for more than 3 months?

Do you frequently drink liquids to aid in swallowing dry food?

Suspicion of Sjögren syndrome from the European League Against Rheumatism SS disease Activity Index questionnaire (at least one domain with a positive item)

| Exclusion criteria |
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| Any of the following conditions |
| History headband neck radiation treatment |
| Active hepatitis C infection (with confirmation by PCR) |
| AIDS |
| Amyloidosis |
| Graft-versus-host disease |
| IgG4-related disease |

| Primary Sjögren syndrome criteria | Points |
|---|---------------|
| Labial salivary gland with focal lymphocytic sialadenitis and focus score of ≥ 1 foci/4 mm ² The histopathologic examination should be performed by a pathologist with expertise in the diagnosis of focal lymphocytic sialadenitis and focus score count, using the protocol described by Daniels et al. | 3 |
| Anti-SSA/Ro positive | 3 |
| Ocular Staining Score ≥ 5 (or van Bijsterveld score ≥ 4) in at least one eye Patients who are normally taking anticholinergic drugs should be evaluated for objective signs of salivary hypofunction and ocular dryness after a sufficient interval without these medications in order for these components to be a valid measure of oral and ocular dryness. Ocular Staining Score described by Witcher et al; van Bijsterveld score described by van Bijsterveld. | 1 |
| Schirmer's test ≤ 5 mm/5 min in at least one eye Patients who are normally taking anticholinergic drugs should be evaluated for objective signs of salivary hypofunction and ocular dryness after a sufficient interval without these medications in order for these components to be a valid measure of oral and ocular dryness. | 1 |
| Unstimulated whole saliva flow rate ≤ 0.1 mL/min Patients who are normally taking anticholinergic drugs should be evaluated for objective signs of salivary hypofunction and ocular dryness after a sufficient interval without these medications in order for these components to be a valid measure of oral and ocular dryness. Unstimulated whole saliva flow rate measurement described by Navazesh and Kumar. | 1 |

REFERENCES:

- Shiboski CH, SHiboski SC, Seror R, et al. 2016 American College of Rheumatology/European League Against Rheumatism classification criteria for primary Sjögren Syndrome. *Arthritis Rheum* 2017, 69:35-45.
- Van Bijsterveld OP. Diagnostic test in the Sicca Syndrome. *Arch Ophtamol* 1969;82:10-14.
- Whitcher JP, Shiboski CH, Shiboski SC, et al. A simplified quantitative method for assessing keratoconjunctivitis sick from the Sjögren Syndrome International Registry. *Am J Ophtalmol* 2010; 149:405-15.
- Daniels TE, Cox D, Shiboski CH, et al. Associations between salivary gland histopathologic diagnosis's and phenotypic features of Sjögren's syndrome among 1726 registry participants. *Arthritis Rheum* 2011;63:2021-30.
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