

- I. Ocular symptoms: a positive response to at least one of three validated 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 three times a day?
- II. Oral symptoms: a positive response to at least one of three validated questions.
 1. Have you had a daily feeling of dry mouth for more than 3 months?
 2. Have you had recurrent or persistently swollen salivary glands as an adult?
 3. Do you frequently drink liquids to aid in swallowing dry foods?
- III. Ocular signs: objective evidence of ocular involvement defined as a positive result to at least one of the following two tests:
 1. Shirmer's I test, performed without anesthesia (≤ 5 mm in 5 min)
 2. Rose Bengal score or other ocular dye score (≥ 4 according to van Bijsterveld's scoring system)
- IV. Histopathology: In minor salivary glands focal lymphocytic sialoadenitis, with a focus score ≥ 1 .
- V. Salivary gland involvement: objective evidence of salivary gland involvement defined by a positive result to at least one of the following diagnostic tests:
 1. Unstimulated whole salivary flow (≤ 1.5 mL in 15 min)
 2. Parotid sialography
 3. Salivary scintigraphy
- VI. Antibodies in the serum to Ro/SS-A or La/SS-B antigens, or both.

^aExclusion criteria: past head and neck radiation treatment, hepatitis C infection, AIDS, preexisting lymphoma, sarcoidosis, graft-versus-host disease, use of anticholinergic drugs. ^bPrimary Sjögren's syndrome: any four of the six items, as long as item IV (histopathology) or VI (serology) is positive; or any three of the four objective-criteria items (III, IV, V, VI). ^cIn patients with a potentially associated disease (e.g., another well-defined connective tissue disease), the presence of item I or item II plus any two from among items III, IV, and V may be considered indicative of secondary Sjögren's syndrome.

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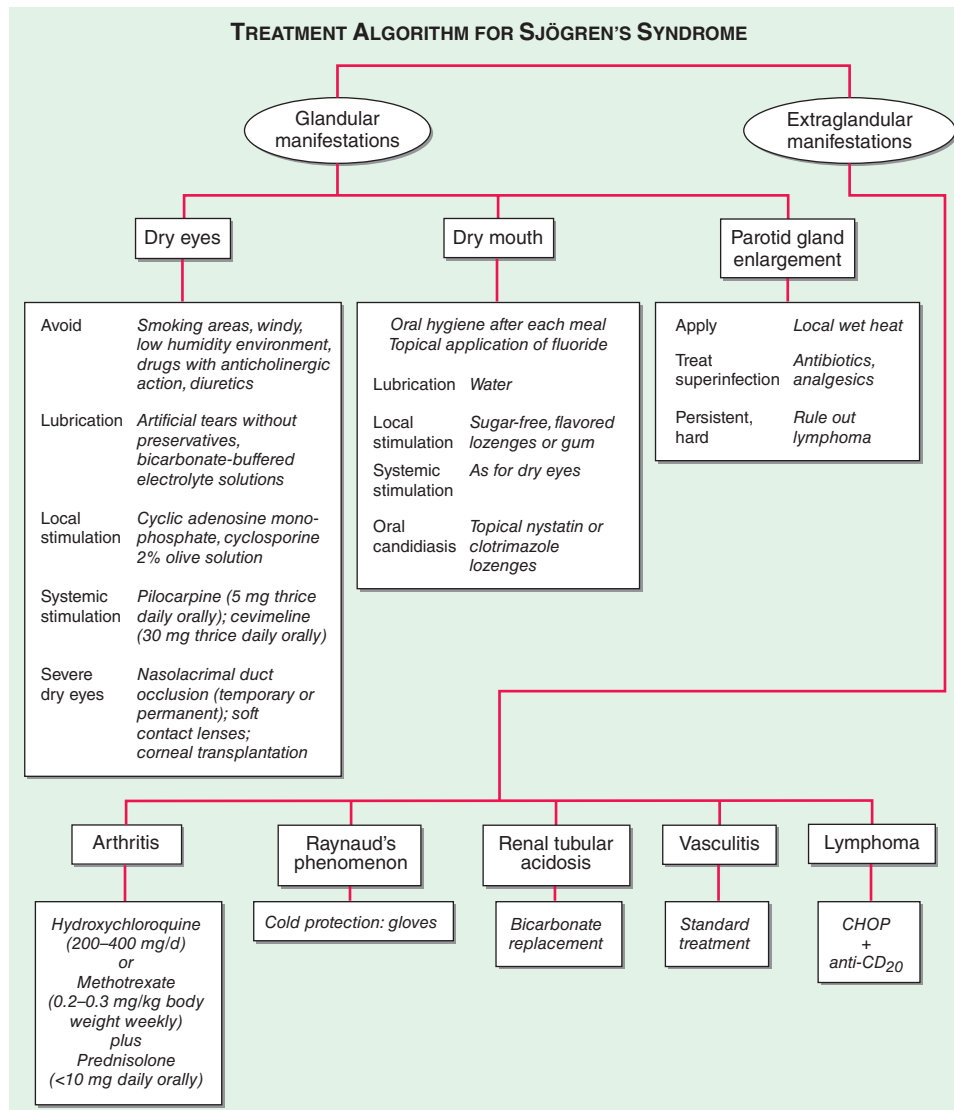


FIGURE 383-1 Treatment algorithm for Sjögren's syndrome. CHOP, cyclophosphamide, adriamycin (hydroxydaunorubicin), vincristine (oncovin), and prednisone.