

- Autoimmune.
- Lymphocytic infiltration of exocrine glands.

Sjogrens syndrome

Disclaimer:
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References:
1. patient.co.uk
2. The British Society for Rheumatology guideline for the management of adults with primary Sjögren's Syndrome
3. Rheumatology consultant HEFT, Walsall Alliance, Mar 2018

Symptoms

- Dry eyes
- Blepharitis
- Corneal lesions
- Dry mouth
- Dental caries
- Oral thrush
- Submandibular gland enlarged
- Parotid gland enlarged

Investigations

- FBC
- ESR
- RF
- ANA
- ENA
- Schirmer test
- USS salivary glands

2o care will diagnose if following apply

- Meets inclusion criteria
- AND
- Does not have any exclusion criteria
- AND
- ACR/EULAR classification criteria score of ≥ 4

- ≥ 1 symptom of ocular or oral dryness
- OR
- Suspicion of Sjögren syndrome from the European League Against Rheumatism SS Disease Activity Index questionnaire (at least 1 domain with a positive item).

- Have you had daily, persistent, troublesome dry eyes for more than 3/12?
- Do you have recurrent sensation of sand or gravel in the eyes?
- Do you use tear substitutes more than 3 times a day?
- Have you had a daily feeling of dry mouth for more than 3/12?
- Do you frequently drink liquids to aid in swallowing dry food?

- Hx of head and neck radiation treatment.
- Active hepatitis C infection (with confirmation by PCR).
- AIDS.
- Sarcoidosis.
- Amyloidosis.
- Graft-versus-host disease.
- IgG4-related disease.

Criteria	Score
- Labial salivary gland with focal lymphocytic sialadenitis and focus score of ≥ 1 foci/4 mm ² .	3
- Anti-SSA/Ro positive.	3
- Ocular Staining Score ≥ 5 (or van Bijsterveld score ≥ 4) in at least 1 eye.	1
- Schirmer's test ≤ 5 mm/5 min in at least 1 eye.	1
- Unstimulated whole saliva flow rate ≤ 0.1 mL/min.	1

Management

- Refer all suspected cases to rheumatologist.
- Steroids, ciclosporin, biologicals etc may be used.
- Simple measures can be advised in 1o care.

Eyes

- Stimulate meibomian gland secretion daily using warm compresses.
- Lubricants (preferably preservative free).
- If fails or severe symptoms refer to ophthalmologist.

- Low viscosity
- Tears Naturale SDU
- Medium (thin) viscosity
- Systane SDU
- Viscotears SDU
- Visidic
- Xailin Fresh 0.5% SDU
- Celluvisc 0.5 or 1% SDU
- Medium (thick) viscosity
- Clinitas 0.4% SDU
- Hylo-Forte
- Hycosan
- Vismed gel 0.3%
- High viscosity
- Lacri-lube
- Xailin Night

Mouth

- Oral hygiene is vital.
- Limit sugar intake.
- Avoid food and drinks other than plain water between meals and from 1hr before bedtime and through the night.
- Regular dentist review (every 6/12).
- Brush teeth at least twice daily (but not immediately after eating) including before bed using a pea sized amount of high fluoride toothpaste and use fluoride-containing oral gel on teeth twice daily.
- Alcohol-free chlorhexidene mouth wash twice daily for maximum of 2/52 every 3/12 to prevent gum disease.
- Use fluoride-containing mouth wash, gel or spray as required for symptomatic relief.
- Chew xylitol-containing sugar-free gum.
- A trial of pilocarpine 5mg OD, increasing stepwise to 5mg QDS if severe dry mouth.

- Simple candida infection (visible white plaques):
= nystatin 1ml five times daily for 1/52. Repeat for 1/52 in 8, if frequent recurrence.
- Erythematous infection (red, raw tongue or oral cavity):
= fluconazole 50mg OD for 10/7.
- Angular cheilitis:
= miconazole topically for 2/52.

Salivary gland

- Consider baseline USS to assess for active inflammation, infection and stones.
- If acute inflammation, in the absence of infection and stones, consider short course of oral prednisolone or intra-muscular Depo-medrone.
- If infection (systemically unwell), prescribe Abx.
- Massaging the glands reduces inflammation in chronically inflamed glands. Start at the ear and massage down and forwards.

Lymphoma

- Counsel patients to report firm, painless glandular swelling that does not settle.
- Investigate suspicious lesions with USS.

Systemic dryness

- A trial of pilocarpine 5mg OD.
- Increase stepwise to max 5mg QDS.

Vaginal dryness

- Topical lubricant.
- Topical oestrogen.

Skin

- Emollients instead of soaps.

Fatigue

- Exercise.

Pregnant

- Consider referral to specialist centre.
- Consider aspirin 75mg OD to improve placental implantation.