



## Sjögren Syndrome

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### WHAT IS SJÖGREN SYNDROME?

Sjögren (show-gren) syndrome is an autoimmune disease where glands that produce tears, saliva, vaginal mucous and other moisture malfunction for some reason. People with Sjögren syndrome often have very dry eyes, mouth and/or vagina, and can have enlarged lymph nodes, neuropathy (nerve inflammation with numbness and tingling) or lung inflammation. Sjögren syndrome may be primary (not associated with other diseases) or secondary (occurring along with another autoimmune disease, such as rheumatoid arthritis, lupus or scleroderma).

Over 20% of people with scleroderma (systemic sclerosis) patients also have Sjögren syndrome, and it's most common in people with limited skin thickening (formerly called CREST syndrome).

### CAUSES

Sjögren syndrome's exact cause is unclear. It happens when lymphocytes, or white blood cells, build up in certain glands and organs. Lymphocytes are not normally found in glands, which points to a faulty immune system as the culprit.

### SYMPTOMS

**Dry Eyes:** "Gritty" or "sandy" feeling in your eyes, mucus build-up in the corners of your eyes, extra sensitivity to bright light (photophobia)

**Dry Mouth:** Reduced saliva that reduces taste and makes chewing food or speaking more difficult. You may crave fluids or need to sip water often. Dry nose and throat reduces sense of smell or causes nosebleeds, hoarseness and dry cough. Parotid glands on your neck just below your ears may enlarge.

**Dry Vagina:** Vaginal dryness and irritation make sexual intercourse uncomfortable

**Other possible symptoms:** Severe fatigue, morning stiffness or swelling of small joints in your hands (arthritis), muscle pain or weakness, and white or blue color changes on fingertips during cold exposure (Raynaud phenomenon)

### DIAGNOSIS

Dryness can have many possible causes, such as medications like antidepressants, anxiety medications, narcotics and antihistamines, as well as aging and menopause. If you experience dryness, see your physician. They will determine if you need to be screened for Sjögren syndrome.

Diagnosis of Sjögren syndrome is based on your symptoms, a physical exam (including a thorough eye exam by an ophthalmologist), blood tests and occasionally, a minor salivary gland biopsy (lip biopsy).

**Eye Tests:** The Schirmer test measures wetness in your eye on a strip of filter paper that's placed inside your lower eyelid. Significant dryness is less than 6 millimeters of wetness after five minutes. Ophthalmologists can perform more sophisticated eye dryness tests.

**Mouth Tests:** Your doctor can measure how much saliva you produce in a few minutes. Your doctor may also refer you to a trained dentist or ENT physician for a salivary gland biopsy. It's a quick procedure where a tissue sample is removed from the tiny gland inside your lower lip to look for high amounts of lymphocytes.

**Laboratory Tests:** Blood tests used to help establish your diagnosis may include anti-SSA and anti-SSB antibodies, rheumatoid factor (RF), antinuclear antibody (ANA), low white blood cell count, high or low total gamma globulin, low

complement (C3 and C4) and high erythrocyte sedimentation rate (ESR).

## COMPLICATIONS

### Eyes:

Severe dryness may cause ulcers on your eye's cornea. You may not be able to wear contact lenses. Scarring, reduced vision, and increased viral and bacterial eye infections are possible too.

### Mouth and Sinuses:

Mouth dryness may increase cavities and gum infections (gingivitis) and lead to tooth loosening. Overgrowth of candida yeast may cause a mouth infection called candidiasis or "thrush" that causes burning sensations and inflammation at the corners of your mouth (angular cheilitis). Sinus infections may be more frequent too.

Your parotid (mumps) glands may slowly, but painlessly swell due to blocked saliva ducts. If gland swelling is rapid and painful with reddened skin and fever, it's a sign of a bacterial infection, so seek emergency treatment.

### Respiratory Tract:

If lymphocytes invade normal tissues, you may develop more frequent respiratory infections like bronchitis and pneumonia. Rarely, accumulated lymphocytes in the lungs may lead to shortness of breath or an abnormal chest X-ray (interstitial fibrosis).

### Nervous System:

Lymphocytes may directly injure nerves in the brain, spinal cord or extremities. If this occurs, you may have disturbances of memory and thought processes, weakness, and abnormal sensation in the lower extremities, bowel and bladder dysfunction, and numbness, tingling, "pins and needles" or burning sensation of the toes and feet. Some of these nerve complications may be permanent.

### Vasculitis:

Rarely, some people with Sjögren syndrome also develop vasculitis, or inflammation of the walls of small blood vessels. The most common sign is a red, spotted leg rash. Some people have numbness, tingling and weakness of the feet and toes.

### Kidney:

In some people, blood and/or protein spills into

in urine and causes mild kidney malfunction. This causes no symptoms, so you should have regular blood and urine tests. Muscle weakness may be caused by excessive potassium loss in your urine that lowers blood potassium.

### Skin:

Skin rashes may occur with sunlight exposure. Rashes may come and go, and look scaly or like round, reddish patches with white centers.

### Pregnancy:

Women with anti-SSA antibody may pass it to their fetus, causing a transient, lupus-like facial rash on the baby after delivery. Another serious complication is permanent fetal heart injury. The fetus may have a very slow heartbeat (heart block), which is often fatal. Women with Sjögren syndrome should consult their rheumatologist and obstetrician before trying to conceive.

### Lymphoma:

Lymphoma (lymphatic cancer) affects about 5% of people with primary Sjögren syndrome, but it is less common in people with secondary systemic sclerosis. Even without pain, if your parotid glands progressively enlarge over a few months, it may be a sign of lymphoma. Your doctor can diagnose lymphoma with a biopsy and other tests. Most lymphomas in Sjögren syndrome respond well to treatment.

## TREATMENT

Treatment for Sjögren syndrome targets symptoms to improve your quality of life:

### Dry Eyes:

- Artificial tear drops, long-acting lubricating pellets in the morning, lubricating ointment at night
- Punctal occlusion, a minor surgery where an ophthalmologist inserts tiny plugs to help retain eye moisture, or ties off the tear ducts with a suture or cauterization

### Dry Mouth:

- Frequent sips of water
- Over-the-counter saliva substitutes or gels, sugar-free chewing gum, Xylitol candies
- Oral candidiasis medications
- Good oral hygiene to prevent cavities: frequent dental cleanings, regular brushing

and flossing, avoiding sugary foods and drinks between meals, fluoride mouth rinses

- Prescription medications like pilocarpine (Salagen®) and cevimeline (Evoxac®)

**Vaginal dryness:**

- Topical vaginal lubricants or moisturizers (not petroleum jelly)

**For other symptoms or complications:**

- Don't smoke and limit alcohol use
- Pace your activities, get adequate exercise and sleep to manage fatigue
- Ibuprofen or naproxen for pain and stiffness
- Hydroxychloroquine (Plaquenil®) for arthritis, skin rash or fatigue
- Immunosuppressants or steroids for serious lung, kidney or nervous system complications, or vasculitis

Oral/dental care: Scleroderma and primary Sjögren syndrome may cause similar symptoms, like reduced mouth opening, fingertip ulcers and finger deformities, that make oral hygiene challenging. If you have scleroderma and/or Sjögren syndrome, see your dentist and periodontist regularly.

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Disclaimer: The information provided is for educational purposes only. Any drugs or treatments mentioned should be discussed with your own physician(s).