

## WHAT IS SJÖGREN’S SYNDROME?

Sjögren’s Syndrome (show-grens) is an autoimmune disease characterized by malfunction of the tear- and saliva-producing glands and the mucous-secreting glands of the vagina. The result is the feeling of dryness of the eyes, mouth, and vagina. Sjögren’s Syndrome may be primary (occurring alone, not associated with other diseases) or secondary (occurring in patients who have another autoimmune disease such as rheumatoid arthritis, systemic lupus erythematosus, or scleroderma). Over 20% of systemic sclerosis patients have Sjögren’s Syndrome, which occurs most commonly in those with limited skin thickening (old term CREST syndrome).

## CAUSES

Sjögren’s Syndrome is caused by accumulation of lymphocytes in affected glands and the ducts which drain these glands. Lymphocytes are normally found in the blood stream, lymph nodes, spleen, and bone marrow. Their appearance in glandular tissues is abnormal, signifying that organization of the immune system has become faulty. When they are present in glandular tissues, lymphocytes interfere with the production and flow of tears, saliva, and vaginal secretions.

## SYMPTOMS

### Dry Eyes

You may have a “gritty” or “sandy” feeling in your eyes. On awakening in the morning you may notice increased thick mucus in the corners of your eyes nearest your nose. Your eyes may be unusually sensitive to bright light (photophobia).

### Dry Mouth

You will produce less saliva, which makes moving food in your mouth and speaking more difficult, and reduces your sense of taste. You will crave water or other liquids. You may not be able to eat a dry cracker without water, or eat a meal without frequent sips of water. You may feel the need to take a bottle of water with you wherever you go. Your nose and throat may also be dry, leading to decreased sense of smell, nosebleeds, hoarseness, and dry cough. Your major saliva producing glands (parotid glands, just below and in front of your ears) may become

painlessly enlarged, giving the appearance of mumps.

### Dry Vagina

You may experience vaginal irritation, making sexual intercourse uncomfortable.

*(Caution to patients: There are many other causes of glandular dryness. Increased age, other diseases of the eyes and mouth, certain medications, and lack of estrogen in the vagina are common reasons for dryness which should be considered by your physicians.)*

### Other Symptoms

You may suffer from fatigue, which can be severe enough to interfere with your lifestyle. You may have stiffness or swelling of the small joints of your hands (arthritis) and other joints. Joint pain and stiffness are typically worse in the morning and improve within one to two hours. Muscle pain or weakness can occur, leading to difficulty arising from a chair or lifting your arms over your head. You could experience white or blue color changes at the tips of your fingers during cold exposure (Raynaud’s phenomenon).

## DIAGNOSIS

This condition is not diagnosed by a blood test. The most important contributors to a correct diagnosis are your medical history and physical examination. In addition, certain tests may be helpful for your physician.

### Eye Tests

The Schirmer test is a screening method to measure how much you are able to wet a strip of filter paper placed inside your lower eyelid. If the paper is wet less than 10 millimeters in five minutes after placement, it is considered a positive test. Reduced tearing should be confirmed by an ophthalmologist, who can perform additional more sophisticated tests and look for other causes of dry eye.

### Mouth Tests

A biopsy of the inside of the lower lip may support the diagnosis. In Sjögren’s Syndrome, a greatly increased number of lymphocytes is seen surrounding the small saliva-producing glands. Injection of dye into the parotid duct (sialogram) can demonstrate characteristic enlargement (dilatation) and distortion of these ducts.

### Laboratory Tests

Only half of Sjögren patients have anti-SSA and/or anti-SSB (Sjögren’s Syndrome A and B) antibodies in their blood. Other tests which may be abnormal include the white blood cell count (low), total gamma globulin level (high), blood C3 and C4 complement



levels (low C4), sedimentation rate (high) and rheumatoid factor (positive).

## COMPLICATIONS

### Eyes

Eye dryness may cause dry spots or ulcers on the cornea, leading to inability to wear contact lenses and in some cases scarring with reduced vision. Affected eyes are more prone to viral and bacterial infections.

### Mouth and Sinuses

An increased number of cavities and infection of the gums (gingivitis) with loosening of the teeth may occur. There is an increased frequency of overgrowth of the common yeast candida, resulting in a mouth infection termed candidiasis (can-di-DYE-ah-sis) or “thrush.” There may be slow and painless enlargement of the parotid (mumps) glands due to blockage of the ducts through which saliva flows from the glands into the mouth. If rapid, painful enlargement of one of these glands occurs, accompanied by redness of the overlying skin and fever, this may indicate a secondary bacterial infection of the gland and is a medical emergency. When lymphocytes interfere with normal handling of infectious agents such as viruses and bacteria, sinus infections become more frequent.

### Respiratory Tract

Again, because lymphocytes invade normal tissues, there is an increased risk of developing infections such as bronchitis and pneumonia. The lung tissue itself may be the site of lymphocyte accumulation, leading to shortness of breath and an abnormal chest x-ray (interstitial fibrosis), but this is uncommon.

### Nervous System

Lymphocytes or their products may directly injure nerves in the brain, spinal cord, or extremities. The results can be 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. These problems may be permanent since the ability of nerves to regenerate is limited.

### Vasculitis

A few patients develop vasculitis (inflammation of the walls of small blood vessels). A red spotted rash on the legs and numbness and tingling of the feet and toes is a clue to this complication. Vasculitis can also affect internal organs such as the heart and intestinal tract and is a serious complication.

### Kidney

Some patients with Sjögren’s Syndrome develop blood and/or protein in the urine and, in rare cases, mild kidney malfunction. There are no urinary symptoms associated with these problems, and thus a periodic urinalysis and blood creatinine level should be checked. Excessive loss of potassium in the urine may lead to a low blood potassium level and muscle weakness.

### Skin

A characteristic rash may appear in skin areas exposed to ultraviolet light or sunlight (photosensitive rash). The rash is most often transient. It may be scaly or appear as circular or oval patches of red skin with a central white area.

### Pregnancy

A woman with Sjögren’s Syndrome and anti-SSA antibody in her blood may pass this antibody across

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the placenta to her developing fetus. One result may be a transient lupus-like facial rash in the infant after delivery. A more serious problem is permanent injury to the fetal heart, leading to a very slow heartbeat (heart block). This problem can occur as early as the third month of pregnancy and is often fatal. For these reasons, women with Sjögren's Syndrome should consult their rheumatologist and obstetrician before becoming pregnant.

### Lymphoma

Rarely, the lymphocytes in Sjögren's patients can become malignant, resulting in lymphoma. This complication typically occurs in patients who have had Sjögren's Syndrome for many years. Patients may complain about excessive fatigue and are found to have multiple painless enlarged lymph nodes or unusually rapid enlargement of a parotid gland. The diagnosis is made by lymph node or parotid gland biopsy. Fortunately, most lymphomas in Sjögren's Syndrome patients respond well to chemotherapy.

### TREATMENT

There is no recognized cure for Sjögren's Syndrome. Therefore, doctors try to treat the symptoms of the disease to minimize their effects on your daily life. The following aids may be recommended:

#### For dry eyes

- Artificial tears, every two to four hours during the day, or a long-acting pellet in the morning and a lubricating ointment at night
- Punctal occlusion, a surgical procedure to retain moisture by preventing the normal flow of tears from the inner corner of the eye into the nose. This can be temporary (by inserting small plastic plugs) or permanent (by tying off the ducts with a suture)

#### For dry mouth

- Sips of water throughout the day or over-the-counter saliva substitutes or gels
- Sugar-free chewing gum or candies to stimulate saliva flow
- Treatment for oral candidiasis
- One of several oral saliva stimulant medications

containing the active ingredient pilocarpine

- Good oral hygiene to prevent cavities: frequent dentist visits for teeth cleaning; brushing and flossing teeth regularly and thoroughly, especially after meals; avoiding sugar-containing foods and drinks between meals; using mouth rinses containing fluoride

#### For vaginal dryness

- Specially designed lubricants, but do not use petroleum jelly (does not moisturize the vaginal lining)

#### For other organs affected

- Common-sense measures: avoid cigarette smoking; pace activities to avoid fatigue; get adequate exercise and sleep
- Aspirin or anti-inflammatory drugs for joint pain and stiffness or muscle pain
- Hydroxychloroquine (Plaquenil®) for arthritis, skin rash and fatigue
- Cortisone or immune system suppressing drugs for more serious problems such as involvement of the lung, kidney, nervous system, or vasculitis

### HOW IS SJÖGREN'S SYNDROME RELATED TO SCLERODERMA?

Over 20 percent of patients with systemic sclerosis and a few with localized scleroderma also have secondary Sjögren's Syndrome. It is more often detected in persons with the limited form of systemic sclerosis. The symptoms and examination findings and methods of diagnosis and treatment in Sjögren's Syndrome secondary to scleroderma are identical to those in primary Sjögren's Syndrome. Special problems encountered by scleroderma patients are reduced mouth opening, finger-tip ulcers, and deformities of the fingers, all of which interfere with maintaining good oral hygiene. Therefore it is particularly important for scleroderma patients to consult their dentists and periodontists to make sure that they use appropriate prophylactic measures.

The Scleroderma Foundation thanks Thomas A. Medsger, Jr., M.D., Gerald P. Rodnan Professor of Medicine, University of Pittsburgh, for his help in preparing this brochure.

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

## OUR THREE-FOLD MISSION IS SUPPORT, EDUCATION AND RESEARCH



**Support:** To help patients and their families cope with scleroderma through mutual support programs, peer counseling, physician referrals, and educational information.

**Education:** To promote public awareness and education through patient and health professional seminars, literature, and publicity campaigns.

**Research:** To stimulate and support research to improve treatment and ultimately find the cause of and cure for scleroderma and related diseases.



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