

# SJÖGREN SYNDROME



**SCLERODERMA  
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# WHAT IS SJÖGREN SYNDROME?

Sjögren Syndrome (pronounced, show-gren) is an autoimmune Rheumatic disease characterized by malfunction of the tear and saliva producing glands and the mucous-secreting glands of the vagina, along with other body organ involvement. The result is the feeling of dryness of the eyes, mouth, and vagina, along with different body system abnormalities such as enlarged lymph nodes, neuropathy (inflammation of nerves leading to numbness and tingling) and lung inflammation. Sjögren 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 scleroderma (systemic sclerosis) patients have Sjögren Syndrome, which occurs most commonly in those with limited skin thickening (old term CREST syndrome).

## CAUSES

There are several theories as to what causes the disease, but in general, Sjögren Syndrome is thought to be caused by accumulation of lymphocytes, a type of white blood cells, in affected glands, and other body organs, which leads to malfunction and development of symptoms, depending on the involved tissue. Lymphocytes are normally found in the blood stream, lymph nodes, spleen, and bone marrow. Their abundant appearance in glandular tissues is abnormal, signifying that organization of the immune system has become faulty.

## 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 may produce less saliva, which makes moving food in your mouth and speaking more difficult and reduces your sense of taste. You may crave water or other liquids. You may not be able to eat a dry cracker or bread 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. It should be emphasized there are many other causes of glandular dryness. There are so many medications that can lead to dryness (antidepressants, anxiety medications, narcotics, antihistamines, etc.).

Additionally, increased age, other diseases of the eyes and mouth, and lack of estrogen in the vagina are common reasons for dryness which should be considered by your physicians. Most patients with dry eyes and mouth DO NOT have Sjogren's syndrome.

## **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 phenomenon).

## **DIAGNOSIS**

The diagnosis of Sjögren Syndrome is based on the presence of suggestive symptoms in the medical history, physical examination (including thorough eye exam by an ophthalmologist), blood tests and occasionally, additional testing such as a minor salivary gland biopsy (lip biopsy).

### **Eye Tests**

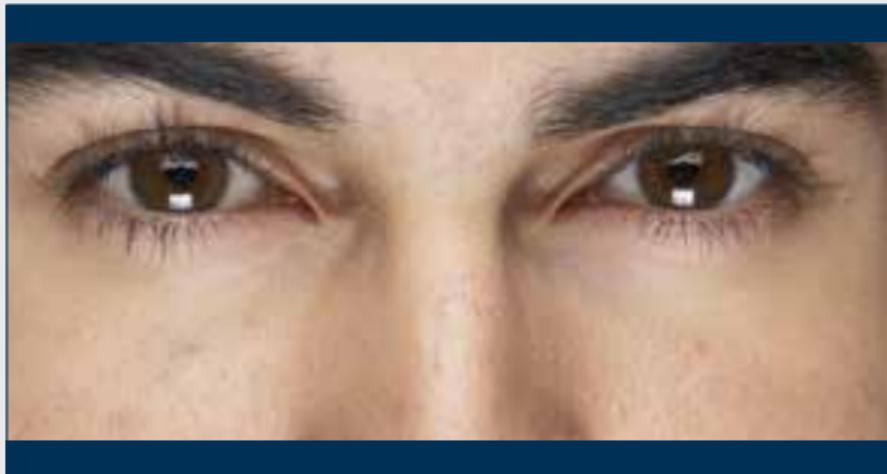
The Schirmer test is a screening method to measure how much you are able to wet a strip of filter paper placed inside your lower eyelid. If the paper is wet less than 6 millimeters in five minutes after placement, it is considered a positive test, indicating significant dryness. An ophthalmologist can perform additional more sophisticated dry eye tests and look for other causes of dry eye.

### **Mouth Tests**

Saliva can be collected in a cup in your doctor's office over a 5 or 15 minute period of time, the amount of saliva can be then weighed to estimate saliva production per minute. A biopsy of the inside of the lower lip may support the diagnosis, particularly in patients who do not exhibit certain antibody in their blood test (called SSA or Sjogren's Syndrome antibody A). In Sjögren Syndrome, a greatly increased number of lymphocytes is seen surrounding the small saliva-producing glands located in the lip. This is a simple and usually safe procedure that can be done in the office and does not require operating room. It is usually performed by a trained dentist or an ENT doctor.

## Laboratory Tests

Anti-SSA is a very important test that should be done in all patients with dry eyes and mouth. Other testing can also be helpful in establishing the diagnosis (anti-SSB antibodies, rheumatoid factor (RF) and antinuclear antibody (ANA). Other tests that may be abnormal include the white blood cell count (low), total gamma globulin level (high or low), complement levels C3 and C4 (low), and erythrocyte sedimentation rate, or ESR (high).



## COMPLICATIONS

### Eyes

Eye dryness may cause 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”. This can be tricky to diagnose as it may only cause burning mouth and inflammation of the corners of the mouth (Angular cheilitis). 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, indicating a bacterial infection of the gland, a medical emergency. When lymphocytes interfere with normal handling of infectious agents (such as viruses and bacteria) by sinus mucosa, 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 is the most common feature of vasculitis in Sjogren’s patients. Numbness, tingling and weakness of the feet and toes are clues to this complication as it affects the nerves. Vasculitis can also affect internal organs such as the heart and intestinal tract and is a serious complication.

## **Kidney**

Some patients may spill blood and/or protein in the urine and develop 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 (Annular erythema), other types of rash can also occur.

## **Pregnancy**

A woman with Sjögren Syndrome and anti-SSA antibody in her blood may pass this antibody across 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 Syndrome should consult their rheumatologist and obstetrician before attempting to become pregnant.

## **Lymphoma**

Occasionally, the lymphocytes in Sjögren patients can become malignant, resulting in lymphoma. This complication affects approximately 5% of primary Sjögren Syndrome patients and is somewhat less common in Sjögren Syndrome patients with secondary systemic sclerosis and typically occurs in those who have had the disease for many years. Lymphoma can develop in the lymph nodes, but also the parotid glands and other tissues such as the skin, lungs, Kidneys and stomach. Recurrent enlargement of the parotid glands is common in Sjögren's patients and usually resolves in 2-4 weeks. A progressive painless enlargement lasting for a few months should alert the doctor about lymphoma development. The diagnosis is made by lymph node or parotid gland biopsy. Fortunately, most lymphomas in Sjögren Syndrome are slow-growing and respond well to chemotherapy.

## **TREATMENT**

As with all autoimmune rheumatic diseases, there is no recognized cure for Sjögren 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 can be helpful. Preservative free tears should be used if needed more than 4 times daily. They may be used every 30 minutes, if needed. Long-acting pellet in the morning and a lubricating ointment at night may be used.
- Punctal occlusion, a minimal surgical procedure, performed by an ophthalmologist, 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 or burning the duct with a probe (cauterization).

### **For dry mouth**

- Sips of water throughout the day or over-the-counter saliva substitutes or gels
- Sugar-free chewing gum or Xylitol-containing 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

- Saliva stimulating medications: Pilocarpine (Salagen<sup>®</sup>) and Cevimeline (Evoxac<sup>®</sup>) are available as prescription drugs

### **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 and excessive alcohol use; pace activities to avoid fatigue; get adequate exercise and sleep
- Non-steroidal anti-inflammatory drugs, such as Ibuprofen and Naproxen, for joint pain and stiffness or muscle pain
- Hydroxychloroquine (Plaquenil<sup>®</sup>) for arthritis, skin rash, fatigue, etc.
- Cortisone or immune system suppressing drugs for more serious problems such as involvement of the lung, kidney, nervous system, or vasculitis

## **HOW IS SJÖGREN SYNDROME RELATED TO SCLERODERMA?**

Over 20 percent of patients with systemic sclerosis and a few with localized scleroderma also have Sjögren Syndrome. It is more often detected in patients with the limited form of systemic sclerosis. The symptoms and examination findings and methods of diagnosis and treatment of Sjögren Syndrome in scleroderma patients are identical to those in primary Sjögren 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 they use appropriate measures to maximize oral hygiene.

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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).

# BECOME A MEMBER OF THE SCLERODERMA FOUNDATION

When you become a member of the Scleroderma Foundation, you support the organization's mission of support, education and research. Your donation helps pay for programs in each of those three areas, including:



- Funding an average of \$1 million in original research grants awarded to investigators annually.
- Helping patients and their families cope with scleroderma through mutual support groups, physician referrals and the National Patient Education Conference.
- Promoting public education of the disease through publications, seminars, patient education events and publicity campaigns.

## **As a member of the Scleroderma Foundation, you will receive:**

- Our quarterly magazine, the "Scleroderma VOICE." The magazine includes updates on the latest scleroderma research and treatments, positive and uplifting stories from patients living with the disease; and tips about how to manage living with scleroderma.
- Information and educational offerings from your local chapter.
- Discounted registration fees to the annual National Patient Education Conference.

**Please consider joining the Scleroderma Foundation today. A membership form is attached on the reverse side of this panel.**

**To become a member of the Scleroderma Foundation, fill out this form, tear at perforation and send with your check or credit card information to:**

Scleroderma Foundation  
Attn: Donations  
300 Rosewood Drive, Suite 105  
Danvers, MA 01923

I would like to become a member and help support the Scleroderma Foundation's efforts to improve the lives of those with scleroderma, and to assist in the search for a cause and cure. Enclosed please find my check (or credit card information) in the amount of \$\_\_\_\_\_.

**Donations of \$25 or more can be acknowledged as members (\$35 or more for international members).**

- I am not interested in member benefits.
- However, I would like to make a contribution in the amount of \$\_\_\_\_\_.

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# 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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