

The Scleroderma Foundation's mission is three-fold:

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

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

☛ To stimulate and support research to improve treatment and ultimately find the cause and cure of scleroderma and related diseases.

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Sjögren's Syndrome in Scleroderma



**SCLERODERMA
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SUPPORT • EDUCATION • RESEARCH

What is Sjögren's Syndrome?

Sjögren's Syndrome is characterized by malfunction of the tear- and saliva-producing glands and the Bartholin's glands in the vagina. The end result is the feeling of dryness of the eyes, mouth, and vagina. Sjögren's (Show-grenz) Syndrome may be primary (not associated with other diseases), or secondary (develop in patients who have rheumatoid arthritis, lupus, or scleroderma).

What causes Sjögren's Syndrome?

Sjögren's Syndrome is caused by accumulation of activated lymphocytes in the affected glands and the ducts, which drain the glands. These lymphocytes interfere with the production and flow of tears, saliva, and vaginal secretions.

What are the symptoms of Sjögren's Syndrome?

Dry Eyes

You may notice a "gritty" or "sandy" feeling in your eyes. On awakening in the morning you may have increased thick mucus visible in the corners of your eyes nearest the 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 can also be dry, leading to decreased sense of smell, nosebleeds, hoarseness, and dry cough.

Dry Vagina

Sjögren's may cause vaginal dryness, which results in irritation and makes sexual intercourse uncomfortable.

(Caution to patients: There are many other causes of the above complaints of 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 may be considered by your physicians.)

Other Symptoms

You may suffer from fatigue, stiffness, and swelling of the small joints of the hands, swollen lymph glands, muscle pain and/or weakness, Raynaud's phenomenon, or numbness, tingling, "pins and needles" feeling, or burning of the toes and feet.

How is Sjögren's Syndrome diagnosed?

In addition to your medical history and physical examination, certain tests may be useful:

Eye Tests

The Schirmer test is a screening test to measure how much you are able to wet a strip of filter paper placed inside your lower eyelid in 5 minutes. More sophisticated tests can be performed by an ophthalmologist.

Mouth Tests

A biopsy of the inside of the lower lip may establish the diagnosis; increased numbers of lymphocytes are seen surrounding the small saliva-producing glands.

Laboratory Tests

Half of Sjögren's patients have anti-SSA and anti-SSB (Sjögren's Syndrome A and B) antibodies in their blood.

Once Sjögren's Syndrome has been diagnosed, your doctor may order other tests to determine the activity of the disease and the extent to which it might have spread beyond glands. Regular physician evaluations should be done to determine the extent of Sjögren's Syndrome, especially any tendency toward lymphoma.

Women with Sjögren's Syndrome and anti-SSA antibody should consult their rheumatologist and obstetrician before becoming pregnant.

What are the complications of Sjögren's Syndrome?

Eyes

Eye dryness may cause dry spots or ulcers on the cornea, leading to scarring with reduced vision and inability to wear contact lenses.

Mouth

An increased number of cavities (dental caries) and infection of the gums (gingivitis) with loosening of the teeth may occur. Overgrowth of the common yeast candida results 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 duct blockage. If abrupt, painful enlargement of these glands with intense redness of the overlying skin and fever are also present, this may indicate a bacterial infection of the gland and is a medical emergency.

Nervous System Involvement

Lymphocytes may directly injure nerves in the brain, spinal cord, or extremities.

Respiratory Tract

There is an increased risk of developing infections of the ears and sinuses, as well as bronchitis and pneumonia. The lung tissue itself may be invaded by lymphocytes.

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 results. Vasculitis can affect other organs and is a serious complication.

Kidney

Some patients with Sjögren's develop blood and/or protein in the urine and, in a few cases, mild kidney failure.

Skin

A characteristic rash may appear in skin areas exposed to ultraviolet light or sunlight (photosensitive rash).

Pregnancy

A woman with Sjögren's and anti-SSA antibody in her blood may pass this antibody across the placenta to her developing fetus. The result may be a transient lupus-like illness called "neonatal lupus." However, there can also be permanent injury to the fetal heart, leading to a very slow heartbeat (heart block) and other heart abnormalities.

Lymphoma

Rarely, the lymphocytes in Sjögren's patients can become malignant, resulting in a lymphoma.

How is Sjögren's Syndrome treated?

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 2 to 4 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 tears from draining out of the eye and into the nose.

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;
- A saliva-stimulant oral medication containing the active ingredient pilocarpine;
- Good oral hygiene to prevent dental caries: frequent checkups and teeth cleaning; brush and floss teeth regularly and thoroughly, especially after meals; avoid sugar-containing foods and drinks between meals; use mouth rinses containing fluoride.

For vaginal dryness:

- Specially designed lubricants, but do not use petroleum jelly.

For other organs affected:

- Common-sense measures: avoid cigarette smoking; pace activities to avoid fatigue; adequate exercise and sleep;
- Aspirin or non-steroidal anti-inflammatory drugs for arthritis 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.

persons with the limited skin-thickening form of systemic sclerosis, a category which includes patients with the CREST syndrome. The symptoms and examination findings and methods of diagnosis and treatment are very similar to those in primary Sjögren's Syndrome.

A special problem for systemic sclerosis patients may be maintaining good oral hygiene because of reduced mouth opening, Raynaud's phenomenon, finger-tip ulcers, and deformities of the fingers.

Summary

Sjögren's Syndrome is rarely a life-threatening disease. However, dryness is likely to last the rest of your life. By using artificial moisturizing methods, you can minimize the symptoms and prevent local complications.

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).

How is Sjögren's Syndrome related to scleroderma?

Over 20% of patients with systemic sclerosis and a few with localized scleroderma suffer from secondary Sjögren's Syndrome. It is more often detected in