

SJÖGREN'S SYNDROME ASSOCIATED WITH SYSTEMIC SCLERODERMA

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Systemic scleroderma is an autoimmune disease that affects the functioning of small blood vessels and leads to excessive scarring. Individuals with scleroderma are at a higher risk of developing other autoimmune diseases, such as Sjögren's syndrome. In this article, we will discuss what Sjögren's syndrome is, and how it is diagnosed, treated, and monitored.

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WHAT IS SJÖGREN'S SYNDROME?

Sjögren's syndrome is a chronic autoimmune disease that causes the immune system to attack the glands responsible for producing saliva (salivary glands) and tears (lacrimal glands). This results in reduced saliva production, leading to a dry mouth and dental problems such as tooth decay. Damage to the lacrimal glands causes excessive dryness in the eyes, with a sensation of having sand in the eyes, which requires the regular use of artificial tears. Although Sjögren's syndrome is commonly known as "dry eyes and dry mouth syndrome," it can also affect other organs such as the lungs, kidneys, lymphatic system, and neurological system.

I HAVE SCLERODERMA. HOW LIKELY IS IT THAT I ALSO HAVE SJÖGREN'S SYNDROME?

It is estimated that about 20% of patients with systemic scleroderma also have Sjögren's syndrome. This is the autoimmune disease that is the most commonly associated with scleroderma. Patients with limited scleroderma or with anti-centromere autoantibodies are more likely to develop Sjögren's syndrome. However, patients may experience dry eyes or mouth due to other causes like medication side effects.



HOW CAN WE DIAGNOSE SJÖGREN'S SYNDROME?

The diagnosis of Sjögren's syndrome is based on a combination of clinical symptoms, blood markers, and disease-specific tests. Blood markers such as anti-SSA, anti-SSB, and anti-Ro52 can be markers of Sjögren's syndrome. An oral medicine specialist can measure the amount of saliva produced to confirm a dysfunction of the salivary glands, and an ophthalmologist can assess dry eyes using the Schirmer test (which measures the amount of tears produced over a five-minute period) and tests to detect any damage to the surface of the eye caused by a lack of tears. A biopsy of the salivary glands in the lip may also be performed to confirm the diagnosis; however, this procedure is rarely required in the context of scleroderma, as the results usually do not change the treatment being administered for Sjögren's syndrome.

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HOW DO WE TREAT SJÖGREN'S SYNDROME?

Treatment for Sjögren's syndrome primarily aims to relieve the symptoms of dry eyes and dry mouth. Some environmental and lifestyle changes are recommended to prevent worsening of these symptoms (see Tables 1 and 2).

For dry eyes, over-the-counter artificial tears in the form of eye drops can be used throughout the day. If eye drops are not effective enough, artificial tears in the form of gels can be tried. Ointments are best used at bedtime to avoid blurred vision. Preservative-free products are preferred and can be applied every 2 to 4 hours, whereas products with preservatives may increase inflammation if used more than 4 times a day. If symptoms persist, your ophthalmologist may recommend more intensive treatments. Omega-3 supplements (2000 to 3000 mg per day) may also help with symptoms.

For dry mouth, sugar-free products such as chewing gum and candy can help stimulate salivation. It's important to use sugar-free products to prevent tooth decay. Long-lasting lozenges can also be inserted inside the mouth. Saliva substitutes (artificial saliva) are available over the counter in mouthwash, spray, or gel form.

Oral medications to stimulate saliva and tear production (pilocarpine - Salagen®; anethole trithione - Sialor®) may be prescribed if the above treatments are not effective. These medications are effective in improving dryness in 60-70% of patients and may also improve dryness of the skin, nose and vagina. These drugs can cause side effects such as hot flashes, sweating, nausea, headaches and increased urination, but can be well tolerated when dosages are adapted and when taken with food. They are not recommended for people with angle-closure glaucoma, severe asthma, or liver dysfunction.

It is strongly recommended that people with Sjögren's syndrome have regular dental checkups every 3 to 6 months because of the increased risk of tooth decay. High fluoride toothpastes, mouthwashes, gels, and varnishes may be prescribed to prevent cavities.



ARE THERE ANY OTHER COMPLICATIONS TO WATCH FOR IN SJÖGREN'S SYNDROME?

People with Sjögren's syndrome should also be monitored for complications involving internal organs, such as the lungs, kidneys, lymphatic system, and neurological system. This includes a medical questionnaire, physical examination, and annual blood tests. It is important to report symptoms such as involuntary weight loss, persistent swelling of the salivary glands (in front of the ears or under the jaw), or swollen lymph nodes to your doctor, as these symptoms can be indicative of lymphoma, a complication that can affect around 5% of people with Sjögren's syndrome.

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TABLE 1

TIPS TO IMPROVE DRY EYES BY MAKING ENVIRONMENTAL AND LIFESTYLE CHANGES

- ▶ Avoid being in low-humidity environments.
- ▶ Use a humidifier in your bedroom to increase moisture in the air.
- ▶ Avoid being in areas with cold air currents, such as those created by air conditioners or fans.
- ▶ Stay away from smoke and dust.
- ▶ Avoid using eye makeup.
- ▶ Take short breaks with eyes closed when reading or using a computer to reduce strain.
- ▶ Apply warm compresses to the eyelids for 5 to 10 minutes at a time, 2 to 4 times a day to increase glandular secretion.
- ▶ Avoid wearing contact lenses. If you must wear contact lenses, use disposable lenses and replace them daily.
- ▶ Wear safety glasses with side shields or moisture chamber glasses that slow the evaporation of tears (such as Ziena® or 7eye® glasses).
- ▶ Consult your doctor about medications that may cause dry eyes and that can be avoided.

* Table adapted from the *Guide de traitement de la xérophtalmie et de la kératoconjonctive sèche chez les patients atteints du syndrome de Sjögren* (A Treatment Guide for Xerophthalmia and Keratoconjunctiva Sicca in Patients with Sjögren's Syndrome), with kind permission from Dr. Alexandra Albert and Dr. Marie-May Collin-Castonguay.

TABLE 2

TIPS TO IMPROVE DRY MOUTH BY MAKING ENVIRONMENTAL AND LIFESTYLE CHANGES

- ▶ Stop smoking, as it can dry and irritate the mouth and increase the risk of developing candidiasis (fungal infection) and periodontitis (gum disease).
- ▶ Stay hydrated by drinking small amounts of water frequently to keep your mouth moist.
- ▶ Gargle with olive oil or coconut oil to soothe your mouth.
- ▶ Avoid acidic or sweet drinks such as carbonated soft drinks, caffeinated drinks (coffee, tea, energy drinks) and alcohol.
- ▶ Avoid acidic foods such as citrus fruits, kiwi, pineapple, strawberries, etc.
- ▶ Avoid very hot drinks or foods.
- ▶ Avoid very dry or hard foods.
- ▶ Avoid very spicy foods.
- ▶ Accompany foods with sauces to make them easier to swallow.
- ▶ Prefer meats that have been simmered or cooked in foil.
- ▶ Prefer fatty meats.
- ▶ Avoid sweet, sticky foods to reduce the risk of tooth decay.
- ▶ Brush your teeth after each meal. If you cannot, rinse with water.
- ▶ Floss daily.
- ▶ Consult your doctor about medications that may cause dry mouth and that can be avoided.

* Table adapted from the *Guide de traitement de la xérostomie chez les patients atteints du syndrome de Sjögren* (A Guide to the Treatment of Xerostomia in Patients with Sjögren's Syndrome) with kind permission from Dr. Alexandra Albert and Dr. Marie-May Collin-Castonguay.

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CONCLUSION

Sjögren's syndrome is an autoimmune disease that can affect people with scleroderma and cause dry mouth and dry eyes. The main focus of treating this syndrome is to relieve the symptoms by optimizing the environment and lifestyle, using local treatments, and prescribing medications when needed. Regular check-ups with specialists in rheumatology, ophthalmology and dentistry are recommended to ensure proper treatment and to detect possible complications.



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