

Sjögren's Syndrome

Sjögren's syndrome (pronounced sho-grins) is a chronic autoimmune disease in which the body's immune system inappropriately attacks one's own tissues, particularly the glands that produce moisture for the eyes, the mouth and elsewhere in the body. This causes the most common symptoms of Sjögren's syndrome, which are dry eyes and dry mouth.

Sjögren's syndrome may occur by itself or in association with another autoimmune condition, such as systemic lupus erythematosus (also called SLE or just lupus), rheumatoid arthritis or scleroderma (a condition that affects the skin and connective tissue).



What are Some of the Symptoms of Sjögren's Syndrome?

Because Sjögren's syndrome often targets the tear and salivary glands, the most common symptoms involve the eyes and mouth. As an autoimmune disease, the syndrome also has the capability to affect many of the body's other organs and tissues, leading to a wide array of symptoms.

Common symptoms include:

- Dryness of the eyes
- Dryness of the mouth
- Difficulty swallowing
- Change in taste
- Frequent cavities in the teeth
- Frequent oral yeast infections
- Swollen salivary glands
- Vaginal dryness,
- Heartburn
- Cough
- Shortness of breath
- Arthritis
- Fatigue

Depending on the organs and tissues affected, other symptoms also may be present.

What Causes Sjögren's Syndrome?

Sjögren's syndrome is thought to be caused by an abnormal reaction of the body's immune system. Lymphocytes are cells in the body's immune system that travel between the blood and the lymphatic system (including the spleen and lymph nodes). They protect the body from infection and cancer. In Sjögren's syndrome, these cells recognize certain tissues in the body, particularly the glands that produce tears and saliva, as "foreign" and attack them, causing inflammation and damage.

A person who develops Sjögren's syndrome probably inherits the risk from one or both parents and is then exposed to some type of environmental trigger (for example, a viral infection). The exact cause is not known. This means that both genetic and non-genetic factors play a role. Genome sequencing of people with Sjögren's syndrome in both the United States and Asia has added greatly to experts' understanding of the genetic factors by identifying at least five different risk-related major gene regions.

Are There Any Other Effects or Concerns?

Many people develop Sjögren's syndrome without any other autoimmune conditions. However, it is common to develop Sjögren's syndrome in association with other autoimmune diseases, such as rheumatoid arthritis and lupus.

Studies have shown that people with Sjögren's syndrome have a higher risk of developing lymphoma. This is a type of cancer.

Also, an antibody (a protein created by the immune system) that may be present in pregnant women with Sjögren's syndrome can cause heart problems in newborns as well as congenital lupus.

How is Sjögren's syndrome diagnosed?

It is often difficult to diagnose Sjögren's syndrome. In fact, it may take years before a definite diagnosis is made. A specialist in autoimmune diseases (known as a rheumatologist) is usually required to establish the diagnosis.

The diagnosis of Sjögren's syndrome is made based on the careful analysis of many factors. A thorough history and physical examination are essential. Also, there are certain laboratory studies that can be helpful when considering the diagnosis. It is important to note that a diagnosis cannot be made based on any specific blood test alone.

In 2016, the American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR) jointly endorsed a new set of criteria for the classification of primary Sjögren's syndrome (SS) derived through international consensus. These criteria are based on testing of the saliva, blood and eyes.

ACR/EULAR classification criteria for primary Sjögren's syndrome	
Item	Weight/Score
Saliva Tests	
Labial salivary gland (salivary glands near the mouth) A focal lymphocytic sialadenitis and focus score of ≥ 1	3

Unstimulated whole saliva flow rate ≤ 0.1 ml/minute	1
Blood Test	
Anti-SSA/SSB (Ro) positive (antibodies to Ro/SSA antigen)	1
Eye Tests	
Ocular staining (measures damage to the surface of the eye) A score ≥ 5 (or van Bijsterveld score ≥ 4) on at least one eye	1
Schirmer's test (measures tear production) ≤ 5 mm/5 minutes on at least one eye	1

The diagnosis of primary Sjögren's syndrome applies to any person who meets the inclusion criteria (has some of these symptoms) and who has a score of four or greater when the weights/scores from the five criteria are totaled.

How is Sjögren's Syndrome Managed?

It is important to recognize that there is no cure for Sjögren's. In addition, because it is a chronic disease, people often require medical therapy for many years.

Goals of therapy vary for each person since treatment is guided by whichever organs are affected.

Dry eyes

Take care of your eyes. Changes can happen to your eyes.

- Artificial tears and ointments can be used to keep the eyes moist.
- Medications like cyclosporine eyedrop/Restasis® can be used as well.
- On occasion, ophthalmologic procedures are needed to maintain eye moisture. See your eye doctor every year.

Dry mouth

Take care of your mouth and teeth. People with Sjögren's syndrome are at increased risk for gum disease and infections.

- See the dentist every six months.
- Brush your teeth for three minutes at least two times a day. Don't forget your gums!
- Floss at least once a day. If away from home, use a toothpick to clean between your teeth.
- When you're done brushing and flossing, rinse your mouth with water.
- Replace your toothbrush every three to four months.
- Artificial saliva tablets can be used to keep your mouth moist.
- Saliva stimulants (pilocarpine/Salagen® and cevimeline/Evoxac®) can be used for dryness of the mouth.
- Treat any nasal symptoms to help prevent mouth breathing.

Fatigue and joint symptoms

For fatigue and joint symptoms, your doctor may prescribe hydroxychloroquine. For joint swelling, your doctor may prescribe medications that are used for rheumatoid arthritis.

Nervous and blood vessel symptoms

When there is evidence of blood vessel damage, nervous system involvement or lung involvement due to Sjögren's syndrome, potent immunosuppression with corticosteroids (such as prednisone) and other immunosuppressants may be required. Your doctor may also use rituximab in severe cases. This is used to control the underlying autoimmune and inflammatory response.

Remember, goals of therapy vary for each person since treatment is guided by whichever organs are affected.

Visit our website for more information about support groups, clinical trials and lifestyle information.

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