

SJÖGREN'S SYNDROME

In the early 20th century, Swedish physician Henrik Sjögren (SHOW-gren) first described a group of women whose chronic arthritis was accompanied by dry eyes and dry mouth. Today rheumatologists know more about the syndrome that is named for Sjögren and—most significantly for patients—can provide advice about how to live with it. In most cases, Sjögren's is an occasionally annoying but treatable condition that should not affect a person's quality of life.

Fast Facts

- Sjögren's syndrome sometimes develops as a complication of another autoimmune disorder.
- Symptoms vary in type and intensity, but most people with Sjögren's are able to live normal lives.
- Although serious complications are rare, regular medical care is important.

What Sjögren's syndrome is

Sjögren's syndrome is an inflammatory disease that can affect many different parts of the body, but most often affects the tear and saliva glands. Patients with this condition may notice irritation, a gritty feeling, or painful burning in the eyes. Dry mouth or difficulty eating dry foods, and swelling of the glands around the face and neck are also common. Some patients experience dryness of other mucous membranes (such as the nasal passages, throat, and vagina) and skin.

“Primary” Sjögren's syndrome occurs in people with no other rheumatologic disease.

“Secondary” Sjögren's occurs in people who do have another rheumatologic disease, most often [systemic lupus erythematosus](#) and [rheumatoid arthritis](#).

Most of the complications of Sjögren's syndrome occur because of decreased tears and saliva. Patients with dry eyes are at increased risk for infections around the eye and may have damage to the cornea. Dry mouth may cause an increase in dental decay, gingivitis (gum inflammation), and oral yeast infections (thrush) that may cause pain and burning. Some patients have episodes of painful swelling in the saliva glands around the face.

Complications in other parts of the body occur rarely in patients with Sjögren's syndrome. Pain and stiffness in the joints with mild swelling may occur in some patients, even in those without [rheumatoid arthritis](#) or [lupus](#). Rashes on the arms and legs related to inflammation in small blood vessels (vasculitis) and inflammation in the lungs, liver, and kidney may occur rarely and be difficult to diagnose. Neurological complications that cause symptoms such as numbness, tingling, and weakness have also been described in some patients.

What causes Sjögren's syndrome

The cause of Sjögren's syndrome is not known, but it is considered an autoimmune disorder. People with this disease have abnormal proteins in their blood suggesting that their immune system, which normally functions to protect the body against cancers and invading infections, is reacting against their own tissue. The decreased production of tears and saliva seen in Sjögren's syndrome occurs when the glands that produce these fluids are damaged by inflammation. Research suggests that genetic factors and possibly viral infections (as yet unidentified) may predispose people to developing this condition.

Who gets Sjögren's syndrome

Between 1 and 4 million Americans, or roughly 1 to 2 percent of the population, have Sjögren's syndrome. This condition can affect people of any age, but symptoms usually appear between the ages of 45 and 55. It affects 10 times as many women as men. About half of affected patients also have [rheumatoid arthritis](#) or other connective tissue diseases, such as [systemic lupus](#).

How Sjögren's syndrome is diagnosed

Diagnosis depends on a combination of symptoms, physical findings, blood tests, and sometimes special studies. Dry eyes and mouth may be early signs of the condition but require further investigation because these symptoms can be caused by many other conditions or medications. Special tests may be used to assess any decrease in tear or saliva production (an example would be the Schirmer test for tear production). An eye examination is helpful in detecting any eye changes seen in Sjögren's. Blood tests can determine the presence of antibodies (immune system cells that help destroy foreign invaders) typical of the disease, including anti-nuclear antibodies (ANA), anti-SSA and SSB antibodies, or rheumatoid factor. Biopsies of saliva glands around the face or under the surface of the inner lip may also sometimes be used to establish a diagnosis.

How Sjögren's syndrome is treated

Treatment is designed to lessen the most bothersome symptoms. Dry eyes usually respond to the use of artificial tears applied regularly during the day or to gels applied at night. Other measures, such as plugging or blocking tear ducts, can be used in more severe cases. Eyedrops that reduce inflammation in the glands around the eyes (cyclosporine- *Restasis*) may be used to increase tear production. Dry mouth can be relieved by drinking water, chewing gum, or using saliva substitutes. Some patients benefit from using prescription medications that stimulate saliva flow, such as pilocarpine (*Salagen*) or cevimuline (*Evoxac*). If patients develop yeast infections, these can be relieved by anti-fungal therapies. The currently available treatments may help relieve some of the dryness but usually some dryness persists.

All patients should receive regular dental care in order to prevent cavities and tooth loss that may occur as a complication of the disorder. Patients with dry eyes should see an ophthalmologist (eye doctor) regularly for signs of damage to the cornea. Patients with excessive redness and pain in the eyes should be evaluated for infections.

[Hydroxychloroquine](#) (*Plaquinel*), an antimalarial drug used in [lupus](#) and [rheumatoid arthritis](#), may be helpful in some patients with Sjögren's syndrome by reducing joint pain and rash experienced by some patients. Patients with rare but serious systemic symptoms, such as fever, rashes, abdominal pain, or lung or kidney problems, may require treatment with corticosteroids such as prednisone (*Deltasone* and others) and/or immunosuppressive agents [methotrexate](#) (*Rheumatrex*), [azathioprine](#) (*Imuran*), [mycophenolate](#) (*Cellcept*), [cyclophosphamide](#) (*Cytosan*).

Broader health impact of Sjögren's syndrome

A vast majority of patients with Sjögren's syndrome remain very healthy, without any serious complications. Patients should be aware that they do face an increased risk for infections in and around the eyes and an increased risk for dental problems—both of which are due to the long-term reduction in tears and saliva.

Rarely, patients may have complications related to inflammation in other body systems, including:

- Joint and muscle pain with fatigue
- Lung problems that may mimic pneumonia
- Abnormal liver and kidney function tests
- Skin rashes related to inflammation of small blood vessels
- Neurologic problems causing weakness and numbness

In a small number of people, Sjögren's syndrome may be associated with lymphoma, a cancer of the lymph glands.

Living with Sjögren's syndrome

People with Sjögren's syndrome are usually able to live normal lives with very few adjustments. When a diagnosis is made, many patients must focus a great deal of attention dealing with dry eyes and dry mouth, but these symptoms tend to subside with time. Any pain or redness in the eyes should be evaluated promptly, as this may signal an infection. To reduce risk for cavities and other dental problems, patients must pay close attention to proper oral hygiene and regular dental care.

Patients should see their physician regularly for general health screening, and should pay close attention to any abnormal swelling in the glands around the face or neck, under the arms, or in the groin areas as this may be a sign of lymphoma.

Points to Remember

Sjögren's syndrome is an autoimmune condition that can occur at any age, but is most common in older women. Many patients develop Sjögren's syndrome as a complication of another autoimmune disease, such as rheumatoid arthritis or lupus. Most of the treatment for Sjögren's syndrome is aimed at relieving symptoms of dry eyes and mouth and preventing and treating long-term complications such as infection and dental disease. Currently available treatments often do not completely eliminate the symptoms of dryness in some patients. Most patients with Sjögren's syndrome remain healthy, but a number of rare complications have been described, including an increased risk for cancer of the lymph glands (lymphoma). Thus, regular medical care and follow up is important for all patients.