

*Questions  
& Answers  
about . . .*

**Sjögren's  
Syndrome**

*National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)  
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## For Your Information

This publication contains information about medications used to treat the health condition discussed here. When this booklet was printed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

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## What Is Sjögren's Syndrome?

Sjögren's (SHOW-grens) syndrome is an autoimmune disease – that is, a disease in which the immune system turns against the body's own cells. Normally, the immune system works to protect us from disease by destroying harmful invading organisms like viruses and bacteria. In the case of Sjögren's syndrome, disease-fighting cells attack various organs, most notably the glands that produce tears and saliva (the lacrimal and salivary glands). Damage to these glands causes a reduction in both the quantity and quality of their secretions. This results in symptoms that include dry eyes and dry mouth. In technical terms, the form of eye dryness associated with Sjögren's syndrome is called keratoconjunctivitis sicca, or KCS, and the symptoms of dry mouth are called xerostomia. Your doctor may use these terms when talking to you about Sjögren's syndrome.

When organs other than the lacrimal and salivary glands are affected, this is known as “extraglandular involvement.” Usually, this occurs in patients with primary Sjögren's syndrome (see page 2). Manifestations include joint inflammation; particular forms of autoimmune thyroid, kidney, liver, lung, and skin disease; and changes in nerve function of the upper or lower limbs. A small proportion of patients may progress to a form of malignant lymphoma.

## Primary Versus Secondary Sjögren's Syndrome

Sjögren's syndrome is classified as either primary or secondary. Both are systemic diseases, meaning they can affect many systems in the body, and they occur with about equal frequency. The primary form causes early and gradually progressive decreased function in the lacrimal and salivary glands, and can include a variety of extraglandular conditions. The secondary form occurs in people who already have another autoimmune connective tissue disease, most commonly rheumatoid arthritis or systemic lupus erythematosus. These people then develop dry eyes or dry mouth.

Some people with Sjögren's syndrome have certain auto antibodies circulating in their blood called *anti-SS-A* and *anti-SS-B*. They are strongly but not exclusively associated with Sjögren's syndrome. Other people with clear evidence of primary Sjögren's syndrome do not have those antibodies.

While people with secondary Sjögren's syndrome generally have less severe ocular and oral problems than those with the primary form, they also have to contend with the effects of their primary disease (i.e., rheumatoid arthritis, systemic lupus erythematosus, etc.)

You might hear Sjögren's syndrome called a rheumatic disease. This means it causes inflammation in joints, muscles, skin, and other organs. Like rheumatoid arthritis and systemic lupus erythematosus, it is also considered one of the autoimmune connective tissue diseases. These conditions affect the framework of the body (joints, muscles, and skin).

## What Are the Symptoms of Sjögren's Syndrome?

Sjögren's syndrome can cause many symptoms. The main ones are:

- **Dry eyes** – Eyes affected by Sjögren's syndrome may burn or itch. Some people say it feels like they have sand in their eyes. Others have trouble with blurry vision, or are bothered by bright light, especially fluorescent lighting.
- **Dry mouth** – Dry mouth may feel chalky or like your mouth is full of cotton. It may be difficult to swallow, speak, or taste. Because you lack the protective effects of saliva, you may develop more dental decay (cavities) and mouth infections.

As noted above, both primary and secondary Sjögren's syndrome can also affect other parts of the body, causing symptoms such as:

- multiple sites of joint and muscle pain
- prolonged dry skin
- skin rashes on the extremities
- chronic dry cough
- vaginal dryness
- numbness or tingling in the extremities
- prolonged fatigue that interferes with daily life.

### **What Causes Dryness in Sjögren's Syndrome?**

Often in the autoimmune attack causing Sjögren's syndrome, white blood cells called lymphocytes initially will target and damage the glands that produce tears and saliva. Although no one knows exactly how this occurs, the damaged glands produce tears and saliva that are diminished in both quantity and quality, leading to the symptoms of dryness of the eyes and mouth.



## Who Gets Sjögren's Syndrome?

Sjögren's syndrome can affect people of either sex and of any age, but most cases occur in women. The average age for onset is late 40s, but in rare cases, Sjögren's syndrome is diagnosed in children.

## What Causes Sjögren's Syndrome?

Researchers think Sjögren's syndrome is caused by a combination of genetic and environmental factors. Several different genes appear to be involved, but scientists are not certain exactly which ones are linked to the disease, since different genes seem to play a role in different people. For example, there is one gene that predisposes Caucasians to the disease. Other genes are linked to Sjögren's syndrome in people of Japanese, Chinese, and African American descent. Simply having one of these genes will not cause a person to develop the disease. Some sort of trigger must activate the immune system.

Scientists think that the trigger may be a viral or bacterial infection. It might work like this: A person who has a Sjögren's-associated gene gets a viral infection. The virus stimulates the immune system to act, but the gene alters the attack, sending fighter cells (lymphocytes) to the glands of the eyes and mouth. Once there, the lymphocytes attack healthy cells, causing the inflammation that damages the

glands and keeps them from working properly. This is an example of autoimmunity. These fighter cells are supposed to die after their attack in a natural process called apoptosis, but in people with Sjögren's syndrome, they continue to attack, causing further damage. Scientists think that resistance to apoptosis may be genetic.

The possibility that the endocrine and nervous systems play a role in the disease is also under investigation.

## **How Is Sjögren's Syndrome Diagnosed?**

Your doctor will diagnose Sjögren's syndrome based on your medical history, a physical exam and results from clinical or laboratory tests. While reviewing your medical history, your doctor will ask questions about your general health, specific symptoms you are experiencing, and medical problems you and your family members have or have had. Your doctor will also ask about any medications you are taking and about lifestyle habits such as smoking or alcohol consumption. During the exam, your doctor will check for clinical signs of Sjögren's syndrome, such as indications of mouth dryness (described on page 7), or signs of other connective tissue diseases.

Depending on what your doctor finds during the history and exam, he or she may want to perform some tests or refer you to a specialist to establish the diagnosis of Sjögren's syndrome and/or to see how severe the problem is and whether the disease is affecting other parts of the body as well.

Some common eye and mouth tests are:

- **Schirmer test** – This test measures tears to see how the lacrimal (tear) glands are working. The doctor puts thin paper strips under the lower eyelids and measures the amount of wetness on the paper after 5 minutes. People with Sjögren's syndrome usually produce less than 8 millimeters of tears.
- **Slit lamp examination** – This test, in which an ophthalmologist uses equipment to magnify and carefully examine the eye, shows how severe the dryness is and whether the outside of the eye is inflamed.
- **Staining with vital dyes (rose bengal or lissamine green)** – These tests show the extent to which dryness has damaged the surface of the eye. To perform one of these tests, the doctor puts a drop of a liquid containing a dye into the lower eyelid. The dye stains the surface of the eye, highlighting any areas of injury, thereby allowing the doctor to see with the slit lamp how much damage has occurred on the surface of the eye.
- **Mouth exam** – The doctor will look outside the mouth for signs of major salivary gland swelling and inside the mouth for signs of dryness. Signs of dry mouth include a dry, sticky lining (called oral mucosa); dental caries (cavities) in characteristic locations; thick saliva, or none at all coming out of the major salivary ducts; redness of

the mouth lining, often associated with a smooth, burning tongue; and sores at the corners of the lips. The doctor might also try to get a sample of saliva, to check its quality and see how much of it the glands are producing.

- **Lip biopsy** – This test is the best way to find out whether dry mouth is caused by Sjögren’s syndrome. To perform this test the doctor removes tiny minor salivary glands from the inside of the lower lip and examines them under the microscope. If the glands contain white blood cells in a particular pattern, the test is positive for the salivary component of Sjögren’s syndrome.

Because there are many causes of dry eyes and dry mouth (including many common medications, other diseases, or previous treatment such as radiation of the head or neck), the doctor needs a thorough history from the patient, and additional tests to see whether other parts of the body are affected. These tests may include:

- **Routine blood tests** – The doctor will take a blood sample to look for levels of different types of blood cells, check blood sugar level, and see how the liver and kidneys are working.
- **Other blood tests** – Various blood tests may be performed to check for antibodies and other immunological substances often found in the blood of people with Sjögren’s syndrome. Antibodies are gamma globulin molecules, called immunoglobulins,

which are important for fighting infection. Everyone has these in their blood, but people with Sjögren's syndrome usually have too many of them. Antibodies that are directed against the individual making them are called auto antibodies. Antibodies that may be present in people with Sjögren's syndrome include the following.

- Immunoglobulins: The three main classes of immunoglobulins can be measured to see if there is a general increase in antibodies.
- Anti-thyroid antibodies: Auto antibodies against the thyroid gland are created when white blood cells (lymphocytes) migrate into the thyroid gland, causing thyroiditis (inflammation of the thyroid), a common problem in people with Sjögren's syndrome.
- Rheumatoid factors (RF): These are auto antibodies commonly found in the blood of people with rheumatoid arthritis as well as in people with Sjögren's syndrome and other autoimmune connective tissue diseases.
- Antinuclear antibodies (ANAs): These are auto antibodies directed at the cells' nuclei. The presence of ANAs in the blood can indicate an autoimmune disorder, including Sjögren's syndrome.
- Sjögren's antibodies, anti-SS-A (or -Ro) and anti-SS-B (or -La): These are specific antinuclear antibodies that occur commonly, but not always, in people with Sjögren's syndrome.

- **Chest x ray** – Sjögren’s syndrome can cause inflammation in the lungs, so the doctor may want to take an x ray to check them.
- **Urinalysis** – The doctor will probably test a sample of your urine to see how well the kidneys are working.

## **What Type of Doctor Diagnoses and Treats Sjögren’s Syndrome?**

Because the symptoms of Sjögren’s syndrome develop gradually and are similar to those of many other diseases, getting a diagnosis can take time; in fact, it may take years to diagnose Sjögren’s syndrome. During those years, depending on the symptoms, a person could see a number of doctors, any of whom could diagnose the disease and be involved in its treatment. Usually, a rheumatologist (a doctor who specializes in diseases of the joints, muscles, and bones) will coordinate treatment among a number of specialists. In a recent survey of a large number of Sjögren’s syndrome patients, the doctors making their first diagnoses were identified as follows, in order of decreasing frequency:

- rheumatologist
- primary care physician/internist
- ophthalmologist (eye specialist)
- otolaryngologist (ear, nose, and throat specialist)

- dentist (oral care specialist)
- neurologist (nerve and brain specialist)
- allergist (allergic disease specialist)
- endocrinologist (endocrine disease specialist)
- oncologist (cancer specialist)

## How Is Sjögren's Syndrome Treated?

Treatment can vary from person to person, depending on what parts of the body are affected. But in all cases, the doctor will help relieve your symptoms, especially dryness. For example, you can use artificial tears to help with dry eyes and saliva stimulants and mouth lubricants for dry mouth. Treatment for both mouth and eye dryness is described in more detail below.

If you have extraglandular involvement (that is, a problem that extends beyond the moisture-producing glands of your eyes and mouth), your doctor – or the appropriate specialist – will also treat those problems. Treatment may include the following:

- nonsteroidal anti-inflammatory drugs, such as ibuprofen (Motrin\*, Advil) for joint or muscle pain

\* Brand names included in this booklet are provided as examples only, and their inclusion does not mean that these products are endorsed by the National Institutes of Health or any other Government agency. Also, if a particular brand name is not mentioned, this does not mean or imply that the product is unsatisfactory.

- corticosteroid medications, such as prednisone, to suppress inflammation that threatens the lungs, kidneys, blood vessels, or nervous system
- immune-modifying drugs such as hydroxychloroquine (Plaquenil), methotrexate (Rheumatrex), and cyclophosphamide (Cytoxan) to control the overactivity of the immune system that, in severe cases, can lead to organ damage.

## What Can I Do About Dry Eyes?

There are many treatments you can try or your doctor can prescribe for dry eyes. Here are some that might help:

- **Artificial tears** – Available by prescription or over the counter under many brand names, these products keep eyes moist by replacing natural tears. Artificial tears come in different thicknesses, so you may have to experiment to find the right one. Some drops contain preservatives that might irritate your eyes. Drops without preservatives usually don't bother the eyes. These drops typically come in single-dose packages to prevent contamination with bacteria.
- **Ointments** – Ointments are thicker than artificial tears. Because they moisturize and protect the eye for several hours, and may blur your vision, they are most effective during sleep.



- **Hydroxypropyl methylcellulose (Lacriserts)** – This is a chemical that lubricates the surface of the eye and slows the evaporation of natural tears. It comes in a small pellet that you put in your lower eyelid. When you add artificial tears, the pellet dissolves and forms a film over your own tears that traps the moisture.
- **Topical Anti-inflammatory Agents** – Topical steroids and cyclosporin A (Restasis) are used if the surface of the eye is inflamed. Topical steroids can increase the pressure in the eye, so your eye pressure must be monitored regularly.
- **Punctal occlusion** – A surgical procedure used to close the tear ducts that drain tears from the eye, helping to keep more natural tears on the eye's surface. For a temporary closure, the doctor inserts collagen or silicone plugs into the ducts. Collagen plugs eventually dissolve, and silicone plugs are “permanent” until they are removed or fall out. For a longer lasting effect, the doctor can use a laser or a heating device called a cautery to seal the ducts.

## General Tips for Eye Care

- Don't use artificial tears that irritate your eyes. If one brand or prescription bothers you, try another. Nonpreserved drops are usually essential for long-term use.
- Practice blinking: You tend to blink less when reading or using the computer. Remember to blink 5 to 6 times a minute.
- Protect your eyes from drafts, breezes, and wind.
- Put humidifiers in the rooms where you spend the most time, including the bedroom, or install a humidifier in your heating and air conditioning unit.
- Don't smoke, and stay out of smoky rooms.
- Apply mascara only to the tips of your lashes so it doesn't get in your eyes. If you use eyeliner or eye shadow, put it only on the skin above your lashes, not on the sensitive skin under your lashes, close to your eyes. Avoid facial creams on the lower lid skin at bedtime if you are awakening with eye irritation.
- Ask your doctor whether any medications that you are taking contribute to dryness. If they do, ask how the dryness can be reduced.

## What Can I Do About Dry Mouth?

There are many remedies for dry mouth. You can try some of them on your own. Your doctor may prescribe others. Here are some many people find useful:

- **Chewing gum and hard candy** – If your salivary glands still produce some saliva, you can stimulate them to make more by chewing gum or sucking on hard candy. However, gum and candy **must** be sugar-free, because dry mouth makes you extremely prone to progressive dental decay (cavities).
- **Water** – Take sips of water or another sugar-free, non-carbonated drink throughout the day to wet your mouth, especially when you are eating or talking. Note that drinking large amounts of liquid throughout the day will not make your mouth any less dry and will make you urinate more often. You should only take small sips of liquid, but not too often. If you sip liquids every few minutes, it may reduce or remove the mucus coating inside your mouth, increasing the feeling of dryness.
- **Lip balm** – You can soothe dry, cracked lips by using oil- or petroleum-based lip balm or lipstick. If your mouth hurts, your doctor may give you medicine in a mouth rinse, ointment, or gel to apply to the sore areas to control pain and inflammation.

- **Saliva substitutes** – If you produce very little saliva or none at all, your doctor might recommend a saliva substitute. These products mimic some of the properties of saliva, which means they make the mouth feel wet. Gel-based saliva substitutes tend to give the longest relief, but as with all saliva products, their effectiveness is limited by the fact that you eventually swallow them. It is best to use these products rather than water when awakening from sleep: They reduce oral symptoms more effectively, and they do not cause excessive urine formation.
- **Prescription medications** – At least two prescription drugs stimulate the salivary glands to produce saliva. These are pilocarpine (Salagen) and cevimeline (Evxac). The effects last for a few hours, and you can take them three or four times a day. However, they are not suitable for everyone, so talk to your doctor about whether they might help you. In trials of these drugs, patients have also experienced some reduction in their dry eye symptoms.

In addition to treatments for dry mouth itself, some people need treatment for its complications. For example, people with dry mouth can easily get a mouth infection from a common yeast called *Candida*. About one-third of people with Sjögren's syndrome experience this infection, which is called *candidiasis*. Most often, it causes red patches to appear, along with a burning sensation. This occurs particularly on the tongue and corners of the lips. Candidiasis is treated with prescription antifungal drugs.

## The Importance of Oral Hygiene

Natural saliva contains substances that rid the mouth of the bacteria that cause dental decay (cavities) and mouth infections, so good oral hygiene is extremely important when you have dry mouth. Here's what you can do to prevent cavities and infections:

- Visit a dentist regularly, at least twice a year, to have your teeth examined and cleaned.
- Rinse your mouth with water several times a day. Don't use mouthwash that contains alcohol, because alcohol is drying.
- Use toothpaste that contains fluoride to gently brush your teeth, gums, and tongue after each meal and before bedtime. Nonfoaming toothpaste is less drying.
- Floss your teeth every day.
- Avoid sugar between meals. That means choosing sugar-free gum, candy, and soda. If you do eat or drink sugary foods, brush your teeth immediately afterward.
- See a dentist right away if you notice anything unusual or have continuous burning or other oral symptoms.
- Ask your dentist whether you need to take fluoride supplements, use a fluoride gel at night, or have a varnish put on your teeth to protect the enamel.

## What Other Parts of the Body Are Involved in Sjögren's Syndrome?

The autoimmune response that causes dry eyes and mouth can cause inflammation throughout the body. People with Sjögren's syndrome may have extraglandular problems, as noted above. Following are examples of extraglandular problems and how they are treated.

**Skin Problems:** People who have Sjögren's syndrome may have dry skin. Some experience only itching, but it can be severe. Others develop cracked, split skin that can easily become infected. Infection is a risk for people with itchy skin, too, particularly if they scratch vigorously. The skin may darken in infected areas, but it returns to normal when the infection clears up and the scratching stops.

To treat dry skin, apply heavy moisturizing creams and ointments three or four times a day to trap moisture in the skin. Lotions, which are lighter than creams and ointments, aren't recommended because they evaporate quickly and can contribute to dry skin. Also, doctors suggest that you take only short showers (less than 5 minutes), use a moisturizing soap, pat your skin almost dry, and then cover it with a cream or ointment. If you take baths, it's a good idea to soak for 10 to 15 minutes to give your skin time to absorb moisture. Having a humidifier in the bedroom can help hydrate your skin, too. If these steps don't help the itching, your doctor might recommend that you use a skin cream or ointment containing steroids.

Some patients who have Sjögren's syndrome, particularly those with lupus, are sensitive to sunlight and can get painful burns from even a little sun exposure, such as through a window. So, if you're sensitive to sunlight, you need to wear sunscreen (at least SPF 15) whenever you go outdoors and try to avoid being in the sun for long periods of time.

**Vaginal Dryness:** Common in postmenopausal women with or without Sjögren's syndrome, vaginal dryness causes painful intercourse. A vaginal moisturizer and a vaginal lubricant can help. Vaginal moisturizers can attract liquid to the dry tissues, helping to maintain moisture, and are designed for regular use. Vaginal lubricants can make intercourse more comfortable, but they don't moisturize, and therefore aren't appropriate for regular use. Oil-based lubricants, such as petroleum jelly, should be avoided, because they trap moisture and can cause sores and hinder the vagina's natural cleaning process. Regular skin creams and ointments relieve dry skin on the outer surface of the vagina (the vulva).

**Lung Problems:** People with Sjögren's syndrome tend to have lung problems caused by inflammation. These conditions include bronchitis (affecting the bronchial tubes) and tracheobronchitis (affecting the windpipe and bronchial tubes). Lung problems are usually caused by white blood cells (lymphocytes) migrating into the lungs and causing a disease called lymphocytic interstitial pneumonitis. Depending on your condition, the doctor may recommend using a humidifier, taking medicines to open the bronchial tubes, or taking corticosteroids to relieve inflammation.

## Protect Your Voice

People with Sjögren's syndrome can develop hoarseness if their vocal cords become inflamed as part of the disease or become irritated from throat dryness or coughing. To prevent further strain on your vocal cords, try not to clear your throat before speaking. Instead, take a sip of water, chew sugar-free gum, or suck on sugar-free candy. Or else make an "h" sound, hum, or laugh to gently bring the vocal cords together so you can get sound out. Clearing your throat does the same thing, but it's hard on the vocal cords, and you want to avoid irritating them further.

Pleurisy, another Sjögren's-related problem, is inflammation of the lining of the lungs. It is treated with corticosteroids and nonsteroidal anti-inflammatory drugs.

**Kidney Problems:** The kidneys filter waste products from the blood and remove them from the body through urine. The most common kidney problem in people with Sjögren's syndrome is interstitial nephritis, or inflammation of the tissue around the kidney's filters, which can occur even before dry eyes and dry mouth. Inflammation of the filters themselves, called glomerulonephritis, is less common. Some people develop renal tubular acidosis, which means they can't get rid of certain acids through urine. The amount of potassium in their blood drops, causing an imbalance in blood chemicals that can affect the heart, muscles, and nerves.



Often, doctors do not treat these problems unless they start to affect kidney function or cause other health problems. However, they keep a close eye on the problem through regular exams, and will prescribe medicines called alkaline agents to balance blood chemicals when necessary. Corticosteroids or immunosuppressants are used to treat more severe cases.

**Nerve Problems:** In some patients with Sjögren's syndrome, the nervous system may be affected. Most often it affects the peripheral nervous system, which contains the nerves that control sensation and movement. Peripheral nervous system changes occurring in Sjögren's syndrome include the following:

- **Peripheral neuropathy** – a problem that occurs when an immune system attack damages nerves in the legs or arms, causing pain, numbness, tingling, and possibly muscle weakness. Sometimes nerves are damaged because inflamed blood vessels cut off their blood supply.
- **Cranial neuropathy** – a problem in which nerve damage causes face pain; loss of feeling in the face, tongue, eyes, ears, or throat; and loss of taste and smell.

Nerve problems are treated with medicines to control pain and, if necessary, with corticosteroids or other drugs to control inflammation.

**Digestive Problems:** Inflammation in the liver can cause hepatitis and cirrhosis (hardening of the liver). Sjögren's syndrome is closely linked to a liver disease called primary biliary cirrhosis (PBC), which causes itching, fatigue, and, eventually, cirrhosis. Many patients with PBC have Sjögren's syndrome. Treatment varies, depending on the problem, but may include pain medicine, anti-inflammatory drugs, steroids, and immunosuppressants.

**Thyroid disorders:** There is a group of autoimmune thyroid disorders that can appear as either the overactive thyroid of Graves' disease or the underactive thyroid of Hashimoto's thyroiditis. Many people with these autoimmune thyroid disorders also have Sjögren's syndrome and many people with Sjögren's syndrome show evidence of thyroid disease.

**Raynaud's phenomenon:** This is a condition in which the blood vessels in the hands, arms, feet, and legs constrict (narrow) when exposed to cold. The result is pain, tingling, and numbness. When vessels constrict, fingers turn white. Shortly after that, they turn blue because of blood that remained in the tissue pools. When new blood rushes in, the fingers turn red. The problem is treated with medicines that dilate blood vessels. Raynaud's phenomenon usually occurs before dryness of the eyes or mouth.

**Vasculitis:** This is an inflammation of the blood vessels, which then become scarred and too narrow for blood to get through to reach the organs. For people with Sjögren's syndrome, vasculitis tends to occur in those who also have Raynaud's phenomenon and lung and liver problems. It can affect different organs at the same time and have gradually developing systemic (*e.g.*, fever, weight loss, arthritis) or localized (*e.g.*, raised rash) clinical presentations.

## Other Autoimmune Connective Tissue Diseases

Patients who have an autoimmune connective tissue diseases other than Sjögren's syndrome (see list below) may subsequently develop the dry eyes and or dry mouth of Sjögren's syndrome. They would then be diagnosed as having *secondary Sjögren's syndrome*, along with their primary connective tissue disease. These other autoimmune connective tissue diseases include:

- **Polymyositis** – an inflammation of the muscles that causes weakness and pain, difficulty moving, and, in some cases, problems breathing and swallowing. If the skin is inflamed too, it's called dermatomyositis. The disease is treated with corticosteroids and immunosuppressants.

- **Rheumatoid arthritis (RA)** – a form of arthritis that is characterized by severe inflammation of the joints. This inflammation can eventually damage the surrounding bones (fingers, hands, knees, etc.). RA can also damage muscles, blood vessels, and major organs. Treatment depends on the severity of the pain and swelling and which body parts are involved. It may include physical therapy, aspirin, rest, nonsteroidal anti-inflammatory agents, steroids, or immunosuppressants.
- **Scleroderma** – a disease in which the body accumulates too much collagen, a protein commonly found in the skin. The result is thick, tight skin and possibly damage to muscles, joints, and internal organs such as the esophagus, intestines, lungs, heart, kidneys, and blood vessels. Treatment is aimed at relieving pain and includes drugs, skin softeners, and physical therapy.
- **Systemic lupus erythematosus (SLE)** – a disease that causes joint and muscle pain, weakness, skin rashes, and, in more severe cases, heart, lung, kidney, and nervous system problems. As with RA, treatment for SLE depends on the symptoms and may include aspirin, rest, steroids, and anti-inflammatory and other drugs, as well as dialysis and high blood pressure medicine.

## Does Sjögren's Syndrome Cause Lymphoma?

A small percentage of people with Sjögren's syndrome develop lymphoma, which involves salivary glands, lymph nodes, the gastrointestinal tract, or the lungs. Persistent enlargement of a major salivary gland should be carefully and regularly observed by your doctor and investigated further if it changes in size in a short period of time. Other symptoms may include the following: (Note that many of these can be symptoms of other problems, including Sjögren's syndrome itself. Nevertheless, it is important to see your doctor if you have any of these symptoms so that any problem can be diagnosed and treated as early as possible.)

- unexplained fever
- night sweats
- constant fatigue
- unexplained weight loss
- itchy skin
- reddened patches on the skin.

If you're worried that you might develop lymphoma, talk to your doctor to learn more about the disease, the symptoms to watch for, any special medical care you might need, and what you can do to relieve your worry.

## Medicines and Dryness

Certain drugs can contribute to eye and mouth dryness. If you take any of the drugs listed below, ask your doctor whether they could be causing symptoms. However, don't stop taking them without asking your doctor – he or she may already have adjusted the dose to help protect you against drying side effects or chosen a drug that's least likely to cause dryness. Drugs that can cause dryness include:

- antihistamines
- decongestants
- diuretics
- some antidiarrhea drugs
- some antipsychotic drugs
- tranquilizers
- some blood pressure medicines
- antidepressants.

## What Research Is Being Done on Sjögren's Syndrome?

Through basic research on the immune system, autoimmunity, genetics, and connective tissue diseases, researchers continue to learn more about Sjögren's syndrome. The hope is that a better understanding of the disease and its causes will lead to better treatments and perhaps even prevention.

Some of the areas of recent research into Sjögren's syndrome include the following:

- **Hormonal factors** – Because Sjögren's syndrome affects mostly women, female reproductive hormones may play a role. Although studies have shown that levels of estrogen and progesterone differ little between women with Sjögren's syndrome and those without, higher levels of prolactin (a hormone that stimulates the production of milk after childbirth and the production of progesterone in the ovary) are found in women with Sjögren's syndrome. Research is also looking at how the disease affects men and women differently.
- **Medication treatment** – Recent studies have shown that cevimeline (Evoxac), is effective at easing dry eyes, as well as dry mouth, and that the immunosuppressive drug cyclophosphamide (Cytoxan) is effective for treating some of the nervous system effects of Sjögren's syndrome. In a mouse model of this disorder, eye drops of an anti-CD4

antibody were effective at promoting moisture. On the other hand, at least two other therapies under investigation for Sjögren's syndrome – the biologic response modifier etanercept (Enbrel) and the mild male hormone dehydroepiandrosterone (DHEA) have not proven to be effective.

- **Prevalence of extraglandular involvement** – Studies have shown that neurological involvement and Sjögren's-related problems with the skin – including alopecia (a condition characterized by hair loss), vitiligo (a condition in which areas of the skin lose their pigment and become white), and vasculitis (a raised rash) – may be more common than previously thought. Studies also indicate that identifying and treating these problems in people with Sjögren's syndrome is an important part of managing the disease. Another study shows that clinical depression is also common among Sjögren's syndrome patients, and may warrant treatment.
- **Predicting lung involvement** – Knowing who is at highest risk of certain complications can enable doctors to identify and treat these problems earlier and more appropriately. One study showed that serum levels of beta-2 microglobulin (a protein made by plasma cells and associated with inflammation) were higher in people who later developed lung problems with primary Sjögren's syndrome.



- **Role of infection** – Doctors believe that infections may trigger Sjögren's syndrome in people genetically predisposed to the disease. Viral infection is under investigation as a possible trigger for Sjögren's syndrome and other autoimmune diseases. Epstein-Barr virus, hepatitis C virus and Coxsackie virus are being studied.
- **Long-term relief for dry mouth** – Gene therapy studies suggest that we may someday be able to insert molecules into salivary glands that will control inflammation and prevent their destruction. Scientists also envision a day when they will be able to transplant salivary glands from one person to another. Development of a safe and effective artificial salivary gland is already underway.

The National Institute of Dental and Craniofacial Research conducts studies to help scientists understand, manage, and treat Sjögren's syndrome. If you think you might like to take part in a clinical trial, speak with your doctor or check [www.clinicaltrials.gov](http://www.clinicaltrials.gov) for a listing of trials for which you may be eligible.

## Where Can People Find More Information About Sjögren's Syndrome?

- **National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS)**  
National Institutes of Health  
AMS Circle  
Bethesda, MD 20892-3675  
Phone: 301-495-4484 or  
877-22-NIAMS (226-4267) (free of charge)  
TTY: 301-565-2966  
Fax: 301-718-6366  
E-mail: [NIAMSInfo@mail.nih.gov](mailto:NIAMSInfo@mail.nih.gov)  
[www.niams.nih.gov](http://www.niams.nih.gov)
- **National Eye Institute**  
National Institutes of Health  
31 Center Drive, MSC 2510  
Bethesda, MD 20892-2510  
Phone: 301-496-5248  
[www.nei.nih.gov](http://www.nei.nih.gov)
- **National Institute of Allergy and Infectious Diseases**  
6610 Rockledge Drive, MSC 6612  
Bethesda, MD 20892-6612  
Phone: 301-496-5717  
Fax: 301-402-0120  
[www.niaid.nih.gov](http://www.niaid.nih.gov)

- **National Institute of Dental and Craniofacial Research**  
National Institutes of Health  
Bethesda, MD 20892–6400  
Phone: 301–496–4261  
E-mail: [nidrinfo@od31.nidr.nih.gov](mailto:nidrinfo@od31.nidr.nih.gov)  
[www.nidcr.nih.gov](http://www.nidcr.nih.gov)
- **National Institute of Dental and Craniofacial Research, Sjögren's Syndrome Clinic**  
Gene Therapy and Therapeutics Branch  
National Institute of Dental and Craniofacial Research  
Warren Grant Magnuson Clinical Center, Building 10,  
Room 1N113  
National Institutes of Health  
9000 Rockville Pike  
Bethesda, MD 20892–1190  
Phone: 301–435–8528  
[www.nidcr.nih.gov/Research/Intramural/GeneTherapy/SjogrensSyndrome/default.htm](http://www.nidcr.nih.gov/Research/Intramural/GeneTherapy/SjogrensSyndrome/default.htm)
- **National Institute of Neurological Disorders and Stroke**  
NIH Neurological Institute  
P.O. Box 5801  
Bethesda, MD 20824  
Phone: 800–352–9424 (free of charge) or 301–496–5751  
[www.ninds.nih.gov](http://www.ninds.nih.gov)
- **American Academy of Dermatology**  
P.O. Box 4014  
Schaumburg, IL 60168–4014  
Phone: 847–330–0230 or 888–462–3376 (free of charge)  
Fax: 847–330–0050  
[www.aad.org](http://www.aad.org)

- **American Academy of Ophthalmology**  
P.O. Box 7424  
San Francisco, CA 94120  
Phone: 415-561-8500  
Fax: 415-561-8533  
[www.aao.org](http://www.aao.org)
  
- **American Association for Dental Research and  
International Association for Dental Research**  
1619 Duke Street  
Alexandria, VA 22314  
Phone: 703-548-0066  
Fax: 703-548-1883  
[www.iadr.com](http://www.iadr.com)
  
- **American College of Rheumatology**  
1800 Century Place, Suite 250  
Atlanta, GA 30345  
Phone: 404-633-3777  
Fax: 404-633-1870  
E-mail: [acr@rheumatology.org](mailto:acr@rheumatology.org)  
[www.rheumatology.org](http://www.rheumatology.org)
  
- **American Dental Association**  
Department of Public Information and Education  
211 East Chicago Avenue  
Chicago, IL 60611  
Phone: 312-440-2500  
Fax: 312-440-2800  
E-mail: [publicinfo@ada.org](mailto:publicinfo@ada.org)  
[www.ada.org](http://www.ada.org)

- **American Autoimmune Related Diseases Association**  
22100 Gratiot Avenue  
E. Detroit, MI 48021-2227  
Phone: 586-776-3900 or 800-598-4668 (free of charge)  
Fax: 586-776-3903  
E-mail: [aarda@aarda.org](mailto:aarda@aarda.org)  
[www.aarda.org](http://www.aarda.org)
- **Arthritis Foundation**  
P.O. Box 7669  
Atlanta, GA 30357-0669  
Phone: 404-872-7100 or 800-568-4045 (free of charge)  
or call your local chapter (listed in the telephone directory)  
[www.arthritis.org](http://www.arthritis.org)
- **Lupus Foundation of America, Inc.**  
2000 L Street, N.W., Suite 710  
Washington, DC 20036  
Phone: 202-349-1155 or 800-558-0121  
Fax: 202-349-1156  
E-mail: [lupusinfo@lupus.org](mailto:lupusinfo@lupus.org)  
[www.lupus.org](http://www.lupus.org)
- **The Myositis Association**  
1233 20th Street, NW, Suite 402  
Washington, DC 20036  
Phone: 202-887-0088 or 800-821-7356 (free of charge)  
Fax: 202-466-8940  
E-mail: [tma@myositis.org](mailto:tma@myositis.org)  
[www.myositis.org](http://www.myositis.org)

- **National Organization for Rare Disorders, Inc.**  
55 Kenosia Avenue  
P.O. Box 1968  
Danbury, CT 06813–1968  
Phone: 203–744–0100 or 800–999–6673 (free of charge)  
Fax: 203–798–2291  
E-mail: orphan@rarediseases.org  
www.rarediseases.org
  
- **Scleroderma Foundation**  
300 Rosewood Drive, Suite #105  
Danvers, MA 01923  
Help Line: 800–722–HOPE (722–4673) (free of charge)  
Phone: 978–463–5843  
Fax: 978–463–5809  
E-mail: sfinfo@scleroderma.org  
www.scleroderma.org
  
- **Scleroderma Research Foundation**  
220 Montgomery Street, Suite 1411  
San Francisco, CA 94104  
Phone: 415–834–9444 or 800–441–CURE (411–2873) (free of charge)  
Fax: 415–834–9177  
E-mail: srfcure@srfcure.org  
www.srfcure.org
  
- **Sjögren’s Syndrome Foundation**  
6707 Democracy Blvd, Suite 325  
Bethesda, MD 20817  
Phone: 301–530–4420, 800–475–6473  
Fax: 301–530–4415  
E-mail: ssf@sjogrens.org  
www.sjogrens.org

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The mission of the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), a part of the Department of Health and Human Services' National Institutes of Health (NIH), is to support research into the causes, treatment, and prevention of arthritis and musculoskeletal and skin diseases; the training of basic and clinical scientists to carry out this research; and the dissemination of information on research progress in these diseases. The National Institute of Arthritis and Musculoskeletal and Skin Diseases Information Clearinghouse is a public service sponsored by the NIAMS that provides health information and information sources. Additional information can be found on the NIAMS Web site at [www.niams.nih.gov](http://www.niams.nih.gov).



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National Institute of Arthritis and  
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