



[Authoritative facts](#) about the skin from the [New Zealand Dermatological Society Incorporated](#).

[Home](#) | [Skin signs of systemic disease](#)

Sjögren syndrome

What is Sjögren syndrome? (SS)

SS is an autoimmune disease that mainly affects secretory glands of the eyes and mouth, stopping them from producing tears and saliva properly. It results in dry eyes (called keratoconjunctivitis sicca), dry mouth (xerostomia) and sometimes arthritis. It develops slowly. When tested, most people with the condition have a positive blood test for anti-Ro antibodies.

Many patients with SS have some features of other autoimmune diseases, such as [systemic lupus erythematosus](#) or [rheumatoid arthritis](#). In this case they are said to have an overlap syndrome or the SS is said to be secondary to the other autoimmune condition. The patient is said to have *primary SS* if other conditions are not present.

Who gets primary SS and how often does it develop?

Primary SS is 9 times more common in females than males. It is especially common in older females. However anyone can develop it. Up to 2% of elderly females have been found to have primary SS.

What is the cause of SS?

The cause of SS is not known. One theory is that viruses (such as [Epstein-Barr virus](#)) may play a role in triggering SS in people whose genetic background predisposes them to SS. However once the condition has been triggered there is an infiltrate of lymphocytes into the secretory glands. This is a slow, low grade inflammatory process that can cause damage to the cells of the glands and thus impairs function of the glands.

Clinical features

Eye problems in SS

Patients usually describe dry or gritty eyes. They may also feel a burning discomfort, notice decreased tear production, increased eye fatigue and sometimes intolerance of bright lights (photophobia). These symptoms may develop because of increased dryness of the eyes causing damage to the lining of the cornea at the front of the eyes.

Dry mouth

This may develop very gradually over a number of years. When first noticed it may only be the sensation of dryness of the mouth. As it progresses patients may notice difficulty swallowing dry food, difficulty speaking for long lengths of time, problems with dentures and sometimes increased tooth decay. [Oral thrush](#) is also more likely to develop.

Skin signs

The most common skin manifestation is [dry or rough skin](#) (xerosis), which is noted by more than half of patients. This may result in itch. Patients with SS also sweat less than people without the disease. Although these symptoms are common, most patients with SS are more bothered by the dry mouth and dry eyes. Vaginal dryness may also occur.

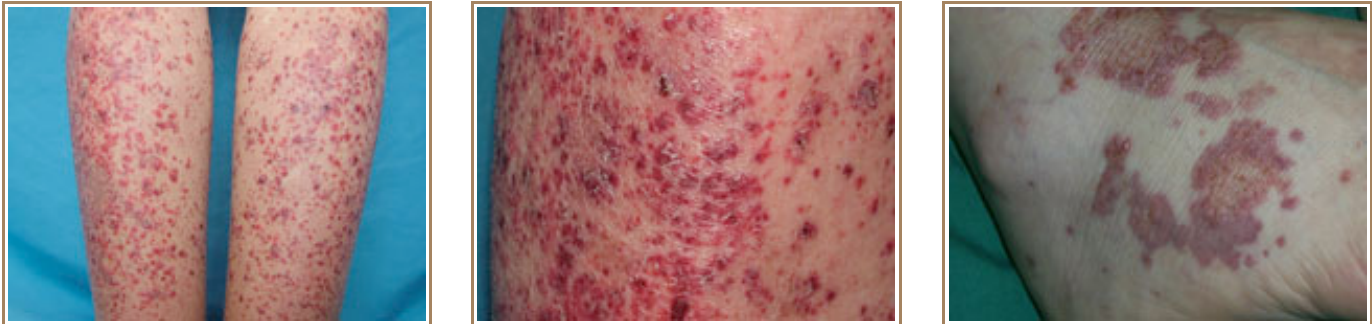
Vasculitis (inflammation of blood vessels) is potentially more serious. This develops in about 10% of patients with primary SS and affects the skin in half of them ([cutaneous vasculitis](#)). Skin lesions usually present as multiple purple or reddish brown spots. The colour is due to bleeding from small inflamed vessels. Vasculitic skin lesions

are most common and prominent on the legs but can develop anywhere. They may present as red flat patches, raised lumps, blisters, ulcers or a raised urticaria-like weals.

Other skin changes that have been reported in Primary SS include:

- [Annular erythema](#) (ring-shaped red patches) has been noted in Japanese patients
- [Livedo reticularis](#)
- [Erythema nodosum](#)
- [Diffuse hair loss](#)
- [Vitiligo](#)
- [Raynaud phenomenon](#)

Sjogren syndrome – cutaneous vasculitis



Involvement of other organs

Gastrointestinal system

Rarely the gastrointestinal system may be affected in SS. The resulting dryness may inflame the lining of the oesophagus (oesophagitis) and the stomach (atrophic gastritis).

Lungs

Although SS may affect the lungs, and is rarely serious. The most common manifestation is a dry cough, due to dryness of the small airways.

Joints

Up to 60% of people with SS will at some stage develop at least one episode of arthralgia (ache in joints) or arthritis (inflammation within the joint). The arthritis does not usually result in erosion of joint structures or in joint deformity.

Kidneys

Involvement of the kidneys is more common if SS is present as part of an overlap syndrome with systemic lupus erythematosus. It can affect the kidneys in various ways including interstitial nephritis, kidney stones or glomerulonephritis.

Vasculitis

Primary SS vasculitis can damage kidneys, lungs, nerves and joints. Fevers and anaemia may also be present. Involvement of internal organs is more likely if the vasculitis is associated with [cryoglobulins](#) (a special form of protein that precipitates at cold temperatures). Some patients with Primary SS and vasculitis have an underlying [B-cell lymphoma](#) (cancer of the lymphatic system).

Long term prognosis

In long term studies, the reduced glandular function in SS has not been observed to improve. If arthritis, Raynaud phenomenon or interstitial nephritis are present early in the course of the illness they can resolve with treatment and time. Worse prognostic factors are the presence of glomerulonephritis, decreased blood levels of C4

complement, vasculitis and cryoglobulinaemia.

Laboratory tests

In people with symptoms that are consistent with SS there are a range of tests that may be done to help confirm the diagnosis.

Schirmer's test.

This measures the wetting of a standardised tear strip applied between the eyeball and the lateral edge of the of the lower eyelid. The test is considered positive if the result is less than 5mm in 5 minutes.

Biopsy of salivary glands in the mouth.

An infiltrate of typical inflammatory cells supports the diagnosis.

Anti-Ro and anti-La antibodies (blood test).

In the correct clinical setting, presence of these antibodies supports a diagnosis of SS.

Other tests that may be performed in special circumstances include the measurement of salivary flow, parotid gland sialography and salivary scintigraphy.

Treatment

The mainstay of treatment for the dry eyes, dry mouth and dry skin is to minimise symptoms by replacing the reduced secretions. There is no cure for SS. Immune suppressive medication is reserved for cases that have internal organ involvement, and are most often used for severe kidney or lung disease.

Dry eyes

Artificial tears and ocular lubricants are most commonly used. In severe cases these may be needed as often as every 30 minutes.

Dry mouth

Increased oral fluid is often taken by the patient as a method to try and relieve symptoms. Other measures include the use of artificial saliva which can come in the form of gels, pastilles or sprays. There is also a medication called pilocarpine which can increase secretion from the damaged glands, although it does not cure the problem or reverse damage to the glands. Pilocarpine can be taken as tablets with each meal and at bedtime to increase the saliva.

Dry skin

[Moisturisers and emollients](#) may be applied as required. Choose a [non-soap cleanser](#). A standard water-based lubricant may be used to relieve dryness of the vagina.

Arthritis

Non steroidal anti-inflammatory drugs are the first line medication for sore joints. Prednisone may be prescribed for severe disease or if symptoms fail to improve with non steroidal treatment.

[Hydroxychloroquine](#) has also been found to be useful in some people.

Vasculitis

[Systemic steroids](#) (prednisone) and immune suppressant medications such as [azathioprine](#) may be necessary.

Related information

References:

- [Sjogren Syndrome](#) - e-medicine dermatology, the online textbook

On DermNet NZ:

- [Cutaneous lupus erythematosus](#)
- [Dry skin](#)

Other websites:

Books about skin diseases:

See the [DermNet NZ bookstore](#)

Author: Dr Scott Barker, Dermatology Registrar, Greenlane Hospital

DermNet does not provide an on-line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

Created 2006. Last updated 10 Dec 2007. © 2007 NZDS. Disclaimer.