



ARTHRITIS FOUNDATION

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SJÖGREN'S SYNDROME

This information leaflet is published by the Arthritis Foundation
as part of our continuing education programme
for all people suffering with arthritis.



The Bone and Joint Decade is a global campaign to improve the quality of life for people with musculoskeletal conditions and to advance understanding treatment of those conditions through research, prevention and education.

WHAT IS SJÖGREN'S SYNDROME?

Sjögren's syndrome is named after Henrik Sjögren (pronounced 'Showgren'), the Swedish ophthalmologist who first described it in 1933. It is an autoimmune disorder of the exocrine glands in which the body's immune system does not work properly. The most common symptoms are dry eyes or a dry mouth (sometimes both together), and feeling very tired and aching.

Although there is no cure for this syndrome, most people can be treated quite easily. A few do have other more serious symptoms. These can also be treated and are described later in this booklet. There are two types of Sjögren's syndrome. Secondary Sjögren's syndrome is caused by another rheumatic illness like rheumatoid arthritis. Primary Sjögren's syndrome is not caused by any underlying rheumatic illness.

This booklet is mainly concerned with primary Sjögren's syndrome. However, the section on diagnosis and treatment will be useful to people with dry eyes or a dry mouth which have been caused by other illnesses.

IS SJÖGREN'S SYNDROME THE SAME AS SICCA SYNDROME?

No. 'Sicca' simply means dryness. Sicca syndrome describes problems of dry eyes and a dry mouth which are not caused by an autoimmune disorder. Sicca syndrome occurs when the tear glands in the mouth which produce saliva are damaged. This damage can be caused by several illnesses, including sarcoidosis, excessive x-ray irradiation and overload disease (haemochromatosis).

HOW COMMON IS PRIMARY SJÖGREN'S SYNDROME?

Primary Sjögren's syndrome is probably the second most common autoimmune rheumatic disorder. It is almost as common as rheumatoid arthritis. It is difficult to work out exact numbers as many people do not seek medical help and simply put up with their symptoms as a bit of a nuisance.

WHO GETS SJÖGGREN'S SYNDROME?

It occurs mostly in women between the ages of 40 and 60. It is much less common in men (1 in 10 of those with Sjögren's syndrome are men), and occurs only rarely in childhood. It affects all races.

WHAT ARE THE MAIN SYMPTOMS?

The most common complaints are:

- dry eyes or a dry mouth (sometimes both dry eyes and a dry mouth)
- feeling very tired and aching.

Many people do not have any other symptoms.



The salivary glands can become swollen in Sjögren's syndrome (as shown by arrows)

HOW DOES SJÖGGREN'S SYNDROME DEVELOP?

As mentioned, Sjögren's syndrome is an autoimmune disorder, so the body's immune system attacks the body's own tissues. In Sjögren's syndrome, the immune system mainly attacks the tear glands in the eye and the salivary glands in the mouth. The immune system sends cells into these glands which cause the glands to swell (to become inflamed)- see Fig. 1. The body also produces antibodies which react with and damage the tissues in the glands. The nerve signals to the glands are also affected and this reduces the amount of saliva produced.

Usually, once the tear and saliva glands have been damaged, the antibodies do not cause any further harm. However, in a few patients, other parts of the body can be affected such as the liver and kidneys. Some joints can become inflamed but the joints are not damaged as they are in rheumatoid arthritis.

WHAT CAUSES SJÖGREN'S SYNDROME?

We are not yet sure how Sjögren's syndrome is caused but it has been linked to several viruses (for example, the Epstein Barr virus, the retroviruses and the HTLV Virus). It is thought that a virus infects the body. When the virus settles within the body tissues, the immune system reacts and this triggers the start of the inflammation.

ARE THERE ANY HEREDITARY INFLUENCES?

There may be some genetic (hereditary) factors which mean that some people find it more difficult to eliminate the infection from their bodies than others. However, it is uncommon for children to inherit the condition from their mother or father.

DOES PREGNANCY AFFECT THE SYNDROME?

There are generally no increased problems during or following pregnancy. However, when mothers have primary Sjögren's syndrome, a few newborn babies may have disurbed heart rhythms. This is because certain antibodies can be passed from mother to baby, but this is very rare.

ARE THERE ANY ALLERGIC REACTIONS?

Sjögren's patients are slightly more likely to develop side-effects to common drugs, eg. antibiotics. We recommend that patients do not take any medications unless they are really necessary.

HOW IS SJÖGREN'S SYNDROME DIAGNOSED?

Sjögren's syndrome can produce a variety of complaints. As a result, patients may need to see various specialists such as their doctor, rheumatologist, eye speicalist, dentist or oral surgeon. After a detailed clinical history and full clinical examination, the main test for Sjögren's syndrome are:

Schirmer's test

A small piece of blotting paper is used to measure tear production.

Slit lamp examination

An eye specialist (ophthalmologist) will place eyedrop dye in the eye (Rose Bengal dye) before examining the

eye with an instrument called a slit lamp. This is a lamp which shines through a very narrow slot, allowing the eye to be examined more effectively.

Measurement of saliva production

This may be measured by spitting into a paper cup over a specific time. More sophisticated investigations include isotope scanning of the salivary glands or injecting contrast dyes into the salivary glands.

Blood test

These are done because two antibodies are found more frequently in primary Sjögren's syndrome than in any other disorder.

Lip Biopsy

One of the tiny salivary glands can be removed very simply from the lower lip without the need for a general anaesthetic. The gland is examined under a microscope to check the diagnosis. This is usually done only if the diagnosis is uncertain based on all the other tests.

ULTRASOUND SCANS AND MAGNETIC RESONANCE IMAGING

Ultrasound scans and *magnetic resonance imaging (MRI)* are sometimes used to help diagnose Sjögren's syndrome.

WHAT IS THE LONG-TERM OUTLOOK IF YOU HAVE SJÖGREN'S SYNDROME?

Unlike some other rheumatic disorder, people with Sjögren's syndrome are not likely to be disabled. The symptoms are annoying and long-lasting but do not usually cause serious problems.

HOW ARE THE MAIN SYMPTOMS OF SJÖGREN'S SYNDROME TREATED?

Table 1 summarises the main symptoms and explains how they can be treated. For all these symptoms, patients should also make sure that they follow guidelines for oral hygiene and reducing dental decay. For example, it is important to follow dental advice and use fluoride gels and remineralisation solution.

TABLE 1. The main symptoms of Sjögren's syndrome and how they can be treated.

Symptom(s)	How they can be treated
Eyes are dry, irritable, and may be sore	Use lubricant eye drops eg natural tears, tear gel, Celluvisc sachets (either a few times a day or up to hourly if necessary).
If strong light are uncomfortable	Wear dark or tinted spectacles if they help. Do not use contact lenses.
Mucous and debris in the eyes cause stickiness	Use mucolytic agents such as acetyl cysteine eye drops
Eyes are very dry, (if there are corneal ulcers or infections these should be referred to an eye specialist at once).	Use preservative-free eye drops, ointments, pseudoplastic drops (Celluvisc). Minor surgery by an eye specialist (Lacriserts punctal occlusion) is advised. Wear special spectacles.
Mouth ulcers cause 'sticky' mouth or throat.	Use diluted chlorhexidine mouthwash. Rinse and gargle with mouthwash (mix 1 teaspoon salt and 1 teaspoon baking powder with 1 litre of water).

TABLE 1. (Continued)

Symptom (s)	How they can be treated
<p>Mouth is very dry.</p> <p>Tastes are altered.</p> <p>Voice is hoarse or weak.</p> <p>Swallowing is difficult (sometimes because the mouth and throat are so dry).</p>	<p>use artificial saliva.</p> <p>Saliva Orthana / Luborant is recommended for patients with their own teeth because they contain fluoride.</p> <p>Glandosane mouth spray can be used by patients with their own teeth but, because it contains no fluoride, care should be taken with dental hygiene and a separate fluoride mouthwash should be used.</p>
	<p>Chew non-sugar chewing gum.</p> <p>Take proper care of dentures.</p>
<p>Mouth is very severely dry causing other problems such as fungal infection, and dry cough*.</p>	<p>Use antifungal treatment such as nystatin.</p> <p>Your doctor may occasionally try bromhexine or pilocarpine.</p>
<p>Dry Cough</p>	<p>Humidifiers and plants with large leaves in rooms may help.</p> <p>Avoid drugs which may aggravate symptoms, eg antihistamines, travel sickness pills and antidepressants.</p>

HOW ARE THE LESS COMMON SYMPTOMS TREATED?

TABLE 2. The less common symptoms of Sjögren's syndrome and how they can be treated.

Symptom (s)	How they can be treated
<p>Pain in the joints (arthralgia).</p> <p>Sometimes there is general aching over the body but also with tender spots (fibromyalgia).</p> <p>Salivary glands are painful.</p>	<p>Take Pain-killers such as paracetamol and / or nonsteroidal antiinflammatory drugs such as ibuprofen.</p> <p>Have an injection of steroid and anaesthetic at isolated tender spots or joints.</p> <p>Ask your doctor for further assessment and advice.</p>
<p>Skin is dry and itchy.</p> <p>Skin is very sensitive to strong sunlight</p>	<p>Avoid strong soaps.</p> <p>Use aqueous creams and emollients, and barrier creams in summer.</p>
<p>Vagina is dry.</p> <p>Intercourse is painful (dyspareunia).</p>	<p>Use lubricants eg K-Y Jelly.</p> <p>Use oestrogen creams.</p> <p>Treat any infection eg thrush.</p>
<p>Feeling extremely tired all the time (lethargy).</p> <p>Joints are inflamed.</p>	<p>Your doctor may prescribe chloroquine or hydroxychloroquine.</p>

TABLE 2. (Continued)

Symptom (s)	How they can be treated
Fever. (These problems are only found in more severe cases).	Corticosteroids such as prednisolone are often prescribed.
Migraine-like headaches.	Take painkillers or specific anti-migraine medications.
Fingers feel cold and turn blue (Raynaud's phenomenon).	Avoid situations which bring on these symptoms. For example, avoid exposure to the cold, do not smoke. Use gloves. Take vasodilators, as prescribed.
Lower abdominal pain and disordered bowel habits (irritable bowel syndrome).	Increase the amount of fibre in the diet. Take antispasmodic drugs eg mebeverine (Colofac)
Liver problems.	These are not very common - mild liver test abnormalities do not require treatment but must be checked regularly by your doctor. Occasionally a specific liver disorder develops called primary biliary cirrhosis. This should be investigated by a specialist.

TABLE 2. (Continued)

Symptom (s)	How they can be treated
Lungs can be affected by fibrosis. This causes breathlessness on exertion, dry cough and occasionally pleurisy causing chest pain.	<p>The lung function must be tested.</p> <p>Fibrosis needs specialist advise.</p> <p>Corticosteroids can be prescribed.</p>
Lymph glands in the neck, armpits or groin are swollen.	<p>Report this to your doctor.</p>
Menopausal symptoms may be aggravated.	<p>Hormone replacement therapy (HRT) can be used. There are no added risks because of Sjögren's syndrome but you should talk to your doctor about the general risks of HRT, which include increased risk of breast cancer and heart disease.</p>
Neuropathy (problems with the <i>peripheral nervous system</i> such as weakness or numbness), <i>vasculitis</i> , or a severe condition overall which is not helped by other treatments	<p>Newer (still experimental) drugs include oral interferon alfa, anti-TNF drugs (etanercept, infliximab and adalimumab), intravenous gammaglobulins and zidovudine.</p>

Glossary

Antibodies – blood proteins which are formed in response to germs, viruses or any other substances which the body sees as foreign or dangerous. The role of antibodies is to attack these foreign substances and make them harmless.

Magnetic resonance imaging (MRI) – a type of scan which uses a strong magnetic field to build up pictures of the inside of the body. It works by detecting water molecules in the body's tissue which give out a particular signal in the magnetic field.

Peripheral nervous system – all parts of the nervous system which are not part of the brain or spinal cord (which form the central nervous system). It includes nerves going to and from the arms, legs, hands, eyes, mouth and other parts of the body.

Ultrasound scan – a type of scan which uses high frequency sound waves to build up images of the inside of the body.

Sarcoidosis – a disorder which causes small fleshy nodules (granulomata) to form in tissues in the body. This can be in the lungs, liver, spleen, skin, salivary glands and other tissues, and the lymph glands are often swollen. The cause is not known.

Vasculitis – inflammation of the walls of blood vessels. This can cause the blood flow to be reduced. Vasculitis can occur on its own (this is called primary vasculitis) or in people who already have an established disease (this is called secondary vasculitis). Secondary vasculitis can happen with a number of different rheumatic disease, including Sjögren's syndrome, rheumatoid arthritis and lupus. There are many different types of vasculitis and there is a separate A.F. booklet on this subject, "Vasculitis".

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