



**BSSA**  
British Sjögren's Syndrome Association

# An Introduction to Sjögren's Syndrome



# About the BSSA

The British Sjögren's Syndrome Association (BSSA) was founded in 1986 as a registered charity, to raise awareness of the disease and support research into its cause and treatment.

A self help organisation with nearly 2300 members, the BSSA is dedicated to providing mutual support and information to individuals affected by this disabling disease. We have regional support groups throughout the UK whose members include sufferers and supporters who work together in helping one another cope with the day-to-day challenges of this debilitating and distressing condition. The BSSA also has a helpline and we award an annual research grant.

## BSSA aims and objectives are:

- To raise awareness of Sjögren's Syndrome
- To educate people about the condition
- To support research into its cause and treatment

## BSSA activities include:

- Providing information and support to those affected by the condition
- Distributing a variety of literature including an informative quarterly newsletter
- Operating a national helpline facility
- Hosting regular meetings throughout the UK
- Linking members through our successful 'make a friend' scheme
- Holding an annual seminar for medical professionals and sufferers, with talks from experts in Sjögren's Syndrome
- Providing a useful and very informative website with an exclusive members only discussion forum

## Become a member

To become a member of the BSSA and enjoy all the benefits we offer, fill in the membership form on this leaflet or visit our website **[www.bssa.uk.net](http://www.bssa.uk.net)**

**UK membership £25.00 per year**

**Overseas membership £30.00 per year**

Sjögren's Syndrome is the UK's second most common autoimmune rheumatic disease, yet the condition remains under recognised and frequently under treated. It does not command a high profile within the medical profession and the general public is largely unaware of the problems faced by sufferers. In reality, Sjögren's Syndrome is a debilitating, distressing and miserable condition.

# What is Sjögren's Syndrome?

Sjögren's Syndrome is an autoimmune rheumatic disease and can be classified as primary, where it occurs alone, or secondary where it occurs in association with another rheumatic disease, most commonly rheumatoid arthritis.

An autoimmune disease is a condition that causes your immune system to react abnormally, attacking the body's healthy cells and tissue instead of fighting infection. In the case of Sjögren's Syndrome, the glands which produce tears, saliva, skin, bowel, vaginal and other secretions are attacked. They develop a form of inflammation and as a result of this these glands stop working leading to dry eyes, dry mouth, dry skin and also dryness of the gastrointestinal tract. Also, dryness of the large bowel can cause symptoms similar to those of irritable bowel syndrome (IBS).

Uncomfortable and unpleasant, the clinical presentation of Sjögren's Syndrome is variable, but typical symptoms include: dry itchy and irritated eyes, joint pain and muscle aches, disabling fatigue, dry mouth and difficulty swallowing, low mood, irritability and impaired concentration.

Sjögren's Syndrome affects women in their 40's and above, although up to 10% of cases occur in men and it is also seen in younger people. It affects one in 2000 women in the UK.

Generalised muscle or joint aching and fatigue can be the most prominent symptoms. The fatigue isn't an ordinary tiredness where you can have a good nights sleep and wake up feeling better, it's a really quite overwhelming fatigue. In the situation where a person's main symptom is tiredness and particularly if the dryness is not reported, the diagnosis may not be made for a long time, if at all. As a result, although the evidence suggests that Sjögren's Syndrome is as common as rheumatoid arthritis, there is a significant proportion of people in whom it is not being correctly diagnosed and people are suffering in silence.



## Dry eyes

In healthy eyes, tears form a moist film over the surface protecting them from dehydration. In Sjögren's Syndrome an immune attack on parts of the eye reduces the volume and quantity of the tears causing the eye to become dry.

A normal eye has three layers:

- The mucin layer which maintains lubrication of the lids during blinking and eye movement.
- The aqueous layer which provides a watery fluid, keeping the surface of the eye moist providing protection from infection through the presence of several proteins. When the eye is open, lacrimal glands produce tears in response to sensory inputs from the surface of the eye and other parts of the body. Emotional events or irritants such as smoke and onion vapour, or the presence of a foreign body in the eye, can stimulate an immediate or more copious amount of tears.
- The lipid layer, which is an oily layer present at the surface of the tear film. This conserves tear water by reducing evaporation. Tear oil is produced by small glands in the eyelids; the presence of oil along the lid margins also prevents tear overflow.

In Sjögren's Syndrome there is a shortage of the watery, lacrimal component of tears (or the second layer) but a disturbance of each of the tear film layers may occur.

The symptoms of dry eye vary from day to day depending on environmental factors and the physical state of health.

In terms of the environment, low humidity encourages water loss and symptoms are worse in air-conditioned rooms, on hot sunny dry days or dry frosty days. Certain occupations can also cause dry eyes such as working at a computer screen. Symptoms are also greatly exacerbated by smoky or dusty atmospheres.

Certain medications\* used in the treatment of allergy, high blood pressure and gastro-intestinal disorders may reduce tear secretion. Antihistamines and some travel sickness pills may reduce tear secretion and should be avoided where possible, by people who have dry eyes.

*\*Please consult your doctor or pharmacist if you are concerned about this.*

## Dry mouth

Saliva is approximately 99% water and has a variety of functions. Not only is it important in maintaining normal oral functions such as taste, chewing and swallowing and ensuring a comfortable mouth, but it also contains substances that help to protect the soft tissues against infection as well as lubricating them. In addition, the flow of saliva helps to reduce the formation of dental plaque and it neutralises the acidity of the mouth when we eat and drink. This function, together with its calcium content helps to protect the teeth against decay.

**A reduction in saliva flow causes dryness of the mouth and is called xerostomia. Not only is this uncomfortable, but it also encourages dental decay and makes patients more vulnerable to soft tissue damage and infection.**

## Causes of dry mouth

### • Non salivary gland causes

Probably the most common cause of dry mouth is medication or dehydration due to infections, poorly controlled diabetes, mouth breathing (especially at night) and acute anxiety.

### • Causes due to salivary glands

Sjögren's Syndrome is probably the commonest cause of xerostomia resulting from disease of the salivary glands themselves.

## Investigation of dry mouth

There are a variety of ways to investigate dry mouth including:

- Salivary flow rates – a simple method of assessing salivary gland function. It is possible to measure stimulated or non-stimulated flow rates, or the flow rate of a single gland or whole salivary flow rate.
- Blood tests – identifying various antibodies (anti Ro and La in particular).
- Minor salivary gland biopsy – removing some of the minor salivary glands under local anaesthetic, usually from the lower lip and examining for a special type of inflammation.
- X-rays and other imaging techniques – there are many techniques now available to identify changes in either the structure or the function of the salivary glands.

## Systemic features

Although dryness of the eyes and mouth are the hallmark of Sjögren's Syndrome, more than one third of patients will have problems affecting other body systems – sometimes referred to as systemic disease.

## Other symptoms can include:

### **Dryness**

- Dryness of the nose and nasal passageways.
- Dry skin.
- Dry and irritating cough caused by the drying up of the secretions in the trachea (windpipe).

### **Fatigue and Joint Symptoms**

- Tiredness and joint pain are common features and may affect more than half of patients with Sjögren's Syndrome.
- Patients can develop an inflammatory arthritis where the joints become hot and swollen.

### **Raynaud's Phenomenon**

- Up to 50% of patients will complain of Raynaud's phenomenon where the blood flow to the hands and feet is affected causing severe coldness and colour changes.

### **Gastrointestinal and liver disease**

- Symptoms similar to those seen in irritable bowel syndrome (IBS) including lower abdominal pain, bloating and change in bowel habit.

- Pancreatitis (inflammation of the pancreas) is a rare complication in Sjögren's Syndrome and usually presents with upper abdominal pain.
- There is also an association between Sjögren's Syndrome and primary biliary cirrhosis.

### **Pregnancy and gynaecological problems**

- Vaginal dryness.
- It seems likely that fertility (i.e. the ability to conceive) is normal in patients with Sjögren's Syndrome but there is an increased risk of miscarriage. The actual risk is difficult to quantify because miscarriage is a common event in any case but at least one study found an increased risk of recurrent miscarriage in women who were positive in certain autoantibodies, which are found in up to 60% of women with Sjögren's Syndrome.

### **Serositis and respiratory disease**

- Serositis (inflammation of the body cavities) may be seen in 5% of patients and presents as sharp, pleuritic chest pain. Rarely Sjögren's Syndrome can cause inflammation of the lung itself, which requires specific investigation and treatment.

### **Renal and urinary tract disease**

- Renal involvement is uncommon but patients have an increased risk of renal stones.
- Interstitial cystitis (inflammation of the bladder wall) may cause very troublesome symptoms, such as urinary frequency.

### **Neurological disease**

Neurological disease is rare. The nervous system is complicated and problems can present in many different ways but do not necessarily imply serious disease.

### **Thyroid**

May affect up to one fifth of patients with Sjögren's Syndrome and usually presents insidiously with an underactive thyroid.

### **Blood test abnormalities**

These include high immunoglobulin levels, low white blood cell count, positive rheumatoid factor, positive ANA and positive Ro and/or La antibodies.

### **Lymphoma**

Lymphomas are cancers of the lymphoid system. The incidence of lymphoma is increased in comparison to the general population but it remains rare and probably affects no more than 1 or 2% of patients with Sjögren's Syndrome.

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Registered charity no. 1101571



# Membership Application

Title: .....

Forename(s): .....

Surname: .....

Date of Birth: .....

Address: .....

.....

Postcode: .....

Contact No: .....

E-mail: .....

Occupation: .....

How did you hear about us? .....

### Payment (please tick):

UK: £25.00       Overseas: £30.00

I would also like to make an additional donation of £.....

### Cheque

To pay by cheque, please complete this application form and send it with your membership fee and, if possible, an additional donation to; **BSSA, PO Box 15040, Birmingham, B31 9DP**. NB: cheques must be made payable to: British Sjögren's Syndrome Association or BSSA.

### Card Payment

We take debit/credit card payments over the phone, please call 0121 478 0222.

### Standing Order

If you would like to pay by standing order, please complete the mandate overleaf and send to the BSSA office.

(NB: Please do not forward the mandate direct to your bank)

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If you are a tax payer, you are eligible for Gift Aid which means any money you give to a charity can be given in a tax efficient way. For each £1 donated to the BSSA, we can claim back 25p from the Government, making your contribution worth £1.25 instead. If you would like us to benefit from your subscription fee/donation, please tick and date the Gift Aid declaration overleaf.



## Standing Order Mandate

(Personal details for BSSA reference)

Name: .....

First line of Address: .....

To: ..... Bank Plc  
..... Branch

Address: .....  
.....

Name of Account to be debited:  
.....

Sort Code: .....

Account No: .....

### Please pay:

HSBC Bank Plc: Harborne, Birmingham  
Sort Code: 40-11-13  
Account No: 51447963  
Beneficiary's Name: British Sjögren's Syndrome Association

### Payment (please tick):

- £25.00 (UK membership only)  
 £30.00 (Overseas membership)  
 I wish to make a further regular donation of £.....  
in addition to my annual subscription fee

Date of first payment: .....

Frequency: **Annually** until further notice

Signature(s): .....

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### Gift Aid declaration (please tick):

- I want the BSSA to treat all subscription fees / donations I have made for the four years prior to this date and all subscription fees / donations I make from the date of this declaration until I notify you otherwise, as Gift Aid donations.