

Sjögren's syndrome

What is Sjögren's Syndrome?

Sjögren's syndrome is an autoimmune disease, where your immune system attacks your healthy cells, especially glands that produce moisture for your body. The disease typically affects glands that make tears in the eyes, and saliva in the mouth causing dry eye or dry mouth or both at the same time. The abnormal immune system causes inflammation and then destruction of the tear glands and salivary glands. These abnormal cells also make antibodies that cause damage to other organs in the body.

This disease can occur by itself (Primary Sjögren's Syndrome), or in association with other autoimmune diseases such as rheumatoid arthritis or systemic lupus erythematosus (secondary Sjögren's Syndrome).

What are the causes of Sjögren's syndrome?

The exact cause of Sjögren's syndrome is not known. It is believed to be a result of genetic factors combined with a previous viral infection that activates the immune system to produce cells that mistakenly attacks its own organs.

Who gets Sjögren's syndrome?

This disease occurs mostly in women between the ages of 40 and 60, although it can affect people at any age.

What are the symptoms of Sjögren's syndrome?

Eyes: dryness, gritty feeling, burning sensation, sensitivity to light.

Mouth: dry sensation, altered taste, difficulty swallowing, recurrent fungal infections, mouth ulcers, and gum infections.

Parotid gland swelling: this causes swelling of the glands on the side of the face, just in front of the ears. Other glands around the face can also be affected.

Skin: very dry skin, and various rashes that cause small red spots on the lower legs or small ulcers.

Vaginal dryness: difficulty with intercourse, and recurrent vaginal or bladder infections.

Lungs: different parts of the lung can be inflamed, causing a dry cough or shortness of breath.

Joints: short lived episodes of joint pain and swelling, which can lead to deformities.

Lymph nodes: swelling of glands other in the neck, armpits or groin, need to be investigated further.

Liver: abnormal liver function tests can be asymptomatic, or a sign of biliary cirrhosis. Your doctor will investigate this further. This is a rare manifestation.

Gut: abdominal pain, diarrhoea, pancreatitis, and difficulty swallowing can also occur. This is not common.

Nerves: numbness tingling and weakness can be a sign of nerve involvement and is common in this disease.

General fatigue: a general feeling of being tired all the time is common and improves with treatment.

Kidney: tubulointerstitial nephritis is a rare finding. It may present with low potassium levels.

Raynaud's: change in colour of the fingers or toes to white, blue and red, especially related to cold, is common in Sjögren's syndrome.

Blood counts: certain cell types are lower than normal in some patients with

Sjögren's, one may get a low white cell count, a low neutrophil count. This may lead to an increased risk of infections.

Cancer: There may be an increased risk of developing a cancer of the lymph nodes called lymphoma.

How is Sjögren's syndrome diagnosed?

Your doctor will take a history and examine you. You will then do some blood tests and may have to go for further tests at the ophthalmologist, or an ENT specialist. The tests required may include:

Blood tests: Antibodies against certain proteins called SSA (Ro) and SSB (La) are measured. These are present in about 50 to 75% of cases.

A Schirmer's test: a small piece of blotting paper is used to measure tear production.

Eye Stains: the ophthalmologist will put special stains in the eye and look for dryness and ulcers.

Lip biopsy: a small punch biopsy of the lower lab is taken by the ENT and sent to the lab for analysis with a microscope. This is only done in certain cases to confirm the diagnosis if the other tests do not help.

What are the treatment options for Sjögren's syndrome?

Eyes: tear replacements, avoid contact lenses, and treat infections with topical antibiotics. Sometimes the ophthalmologist will opt to plug the tear duct that leads from the eye into the nose. For severe disease, you may need to start immunosuppressive treatment.

Mouth: sipping water throughout the day, chewing sugar free gum, or using saliva substitutes may relieve dry mouth. Some patients benefit from using prescription medications that stimulate saliva flow. If there is a fungal infection in the mouth your doctor will prescribe antifungal therapy. Humidifiers and nasal saline irrigation may improve nasal dryness. Using a chlorhexidine mouthwash also helps to decrease tooth and gum infections.

Skin: use aqueous creams, emollient creams, and avoid strong soaps.

Vagina: use lubrication when needed.

Joint pains, fatigue, fevers: Your doctor may prescribe anti-inflammatories, cortisone, or Chloroquine.

Other organ involvement is not common but when it does occur, your physician or rheumatologist will consider using more aggressive immunosuppressive therapy such as methotrexate, azathioprine, cyclosporin, cyclophosphamide, mycophenolate mofetil, or a biologic called rituximab.

What is the long-term outlook of this disease?

As there is no cure for Sjögren's syndrome, it is natural to feel scared, frustrated, sad and sometimes angry.

The majority of patients with Sjögren's syndrome remain very healthy, without any serious complications. Patients should know that they face an increased risk for infections in and around the eyes and an increased risk for dental problems due to the long-term decrease in tears and saliva.

Rarely, patients may have complications related to inflammation in other body systems. If this does occur, your rheumatologist will treat it accordingly.

What else can you do to help your condition?

Stay physically active, eat a healthy diet, stop smoking and reduce stress to help your overall health and wellbeing.

Useful websites:

www.arthritis.org.za

www.sjogrens.org

www.uptodate.com

www.mayoclinic.com

www.rheuminfo.com