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Sjögren syndrome

Sjögren syndrome is an autoimmune disorder in which the glands that produce tears and saliva are destroyed. This causes dry mouth and dry eyes. The condition may affect other parts of the body, including the kidneys and lungs.

Causes

The cause of Sjögren syndrome is unknown. It is an autoimmune disorder. This means the body attacks healthy tissue by mistake. The syndrome occurs most often in women ages 40 to 50. It is rare in children.

Primary Sjögren syndrome is defined as dry eyes and dry mouth without another autoimmune disorder.

Secondary Sjögren syndrome occurs along with another autoimmune disorder, such as:

- Rheumatoid arthritis (RA)
- Systemic lupus erythematosus
- Scleroderma
- Polymyositis
- Hepatitis C can affect the salivary glands and look like Sjögren syndrome

Symptoms

Dry eyes and dry mouth are the most common symptoms of this syndrome.

Eye symptoms:

- Itching eyes
- Feeling that something is in the eye

Mouth and throat symptoms:

- Difficulty swallowing or eating dry foods
- Loss of sense of taste
- Problems speaking
- Thick or stringy saliva
- Mouth sores or pain
- Teeth decay and gum inflammation
- Hoarseness

Other symptoms may include:

- Fatigue
- Fever
- Change in the color of hands or feet with cold exposure (Raynaud phenomenon)
- Joint pain or joint swelling
- Swollen glands
- Skin rash
- Numbness and pain due to neuropathy
- Cough and shortness of breath due to lung disease
- Irregular heartbeat
- Nausea and heartburn
- Vaginal dryness or painful urination

Exams and Tests

A complete physical exam will be done. The exam reveals dry eyes and dry mouth. There may be mouth sores, decayed teeth or gum inflammation. This occurs because of the mouth dryness. Your health care provider will look in your mouth for fungus infection (candida). Skin may show a rash, the lung exam may be abnormal, the abdomen will be palpated for liver enlargement. The joints will be examined for arthritis. The neuro exam will look for deficits.

You may have the following tests done:

- Complete blood chemistry with liver enzymes
- Complete blood count
- Urinalysis
- Antinuclear antibodies (ANA) test
- Rheumatoid factor (possible)
- Test for cryoglobulins

- Complement levels
- Protein electrophoresis
- Test for hepatitis C
- Thyroid tests
- Schirmer test of tear production
- Salivary gland biopsy
- Skin biopsy if a rash is present
- Examination of the eyes
- Chest x-ray

Treatment

The goal is to relieve symptoms.

- Dry eyes may be treated with artificial tears, eye-lubricating ointments, or cyclosporine liquid.
- If Candida is present, it may be treated with sugar-free miconazole or nystatin preparations.
- Tiny plugs can be placed in the tear drainage ducts to help the tears stay on the surface of the eye.

Disease-modifying antirheumatic drugs (DMARDs) similar to those used for RA may improve the symptoms of Sjögren syndrome. These include tumor necrosis factor (TNF) inhibiting drugs such as Enbrel, Humira or Remicaide.

Some things you can do to ease symptoms include:

- Sip water throughout the day
- Chew sugarless gum
- Avoid medicines that can cause mouth dryness, such as antihistamines and decongestants
- Avoid alcohol

Talk with your dentist about:

- Mouth rinses to replace minerals in your teeth
- Saliva substitutes
- Drugs that help your salivary glands make more saliva

To prevent dental decay caused by mouth dryness:

- Brush and floss your teeth often
- Visit the dentist for regular checkups and cleanings

Outlook (Prognosis)

The disease is most often not life-threatening. The outcome depends on what other diseases you have.

There is a higher risk for lymphoma when Sjögren syndrome has been very active for a long time.

Possible Complications

Complications may include:

- Damage to the eye
- Dental cavities
- Kidney failure (rare)
- Lymphoma
- Pulmonary disease
- Vasculitis (rare)
- Neuropathy
- Bladder inflammation

When to Contact a Medical Professional

Call your provider if you develop symptoms of Sjögren syndrome.

Alternative Names

Xerostomia – Sjögren syndrome; Keratoconjunctivitis sicca – Sjögren; Sicca syndrome

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