

# Stay Alert for Atypical Sjögren's Signs, Symptoms

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FORT LAUDERDALE, FLA. — Sjögren's syndrome is the second most common autoimmune disorder that affects the musculoskeletal system, and yet the average time to diagnosis is 6 years, said Yvonne Sherrer, M.D., said at a meeting sponsored by the Sjögren's Syndrome Foundation.

Until more reliable biomarkers for the syndrome are identified, expediting the diagnosis may require physicians to have a higher level of suspicion for some of the less common manifestations of Sjögren's, said Dr. Sherrer, medical director and director of clinical research at the Centre for Rheumatology, Immunology, and Arthritis in Fort Lauderdale.

Although the cause of the Sjögren's is still unknown, researchers suspect that a combination of genetic, environmental, and hormonal factors contribute to predisposition for the disease. Indeed, for every male with the syndrome, an estimated nine women are affected, underscoring the relevance of hormonal influences.

Inflammation of the exocrine glands, the common denominator of Sjögren's syndrome, most obviously affects the eyes, mouth, and vagina, Dr. Sherrer said.

Typically, Sjögren's occurs in the context of a previously diagnosed autoimmune disorder, such as lupus, rheumatoid arthritis, or scleroderma.

The atypical patients with primary Sjögren's "are our most challenging patients, because they don't have an accompanying

autoimmune disease," Dr. Sherrer said. These patients generally present with neuropathy, accelerated dental caries, salivary gland swelling, joint pain without overt arthritis or myositis, and corneal melt. The current international criteria for diagnosing Sjögren's require that patients demonstrate some objective evidence of autoimmunity, in addition to other symptoms. (See box.)

However, the following less typical symptoms may also warrant suspecting Sjögren's syndrome:

► **Ocular.** In addition to extreme dry eyes, patients may suffer from conjunctivitis, keratitis, blepharitis, ulcerations, and perforations.

► **Ears, Nose, and Throat.** Tracheal dryness causes a chronic dry cough in some patients. Nosebleeds, otitis, and sinusitis can be recurring problems.

► **Oral.** Severe dry mouth can cause swallowing problems, which may lead to malnourishment and excessive weight loss. Patients may also have accelerated caries, loss of dentition, and malfunctioning dentures.

► **Dermatologic/Vascular.** Skin rashes are common, and skin eruptions and purpura may occur. Raynaud's phenomenon is a typical vascular manifestation. Vasculitis is always a concern in Sjögren's patients, but symptoms vary depending on the location of the inflammation in the body.

► **Gastrointestinal.** Patients may suffer from esophageal dysmotility. In severe cases, they are at increased risk for pancreatitis, hepatitis, or atrophic gastritis.

► **Hematologic.** Anemia, blood dyscrasias, and cryoglobulinemias are rare but may occur. In addition, Sjögren's patients are at increased risk for lymphoma.

► **Pulmonary.** Lung involvement and coronary involvement are rare but can develop due to dryness of bronchial tubes. Other potential manifestations include bronchitis, bronchitis obliterans-organized pneumonia, and interstitial fibrosis.

► **Neurologic.** Neuropathies tend to be less symmetrical in Sjögren's patients, compared with other conditions. Central nervous system disorders might manifest as changes in cognitive function or as seizures.

► **Renal.** Renal involvement is rare, but when it occurs, it is usually interstitial tube involvement and is more likely to be chron-

ic, compared with highly progressive kidney involvement associated with lupus.

► **Musculoskeletal.** More often than not, patients with Sjögren's have arthralgia, rather than arthritis, but secondary Sjögren's patients may have concurrent arthritis or myositis.

► **Other.** Fibromyalgia occurs in 20%-30% of Sjögren's patients. Sleep disorders, often due to dryness-related discomfort, are common and may contribute to the fibromyalgia. Depression is a common comorbidity, as it is in patients with other autoimmune disease. ■

## Recommended Diagnostic Criteria

The diagnosis of primary Sjögren's syndrome requires that patients meet at least four of the following six criteria:

**1. The patient must have at least one of three ocular symptoms:**

- Dry eyes for less than 3 months.
- Need to use artificial tears more than three times daily.
- Sensation of a foreign body in the eye.

**2. The patient must have at least of three oral symptoms:**

- Persistent dry mouth for more than 3 months.
- Swollen salivary glands.
- Need to add extra liquid to the mouth in order to swallow.

**3. The patient must have at least one of two ocular signs:**

- Unanesthetized Schirmer's test result of 5 mm/5 minutes or less in both eyes.
- Positive vital dye staining.

**4. The patient must have at least one of three signs of poor salivary gland function:**

- Abnormal salivary scintigraphy.
- Abnormal parotid sialography.
- Unstimulated salivary flow rate of 0.1 mL/minute or less.

**5. Positive lip biopsy.**

**6. Positive anti-SSA or anti-SSB tests.**

Source: "The New Sjögren's Syndrome Handbook" (New York: Oxford University Press, 2005)

## Manage Dry Eyes Multiple Ways

FORT LAUDERDALE, FLA. — Optimal management of dry eyes associated with Sjögren's syndrome isn't likely to involve just one solution.

Artificial tears come in assorted formulas, and patients can best manage their eye conditions by recognizing that a single product isn't likely to meet their various needs, William B. Trattler, M.D., said at a meeting sponsored by the Sjögren's Syndrome Foundation.

Patients who complain of burning and irritation from artificial tears should try a preservative-free formula, said Dr. Trattler, an ophthalmologist in private practice in Miami, with a special interest in external eye disease.

Preservative-free drops are typically sold as single-use vials, but a vial can be used in the morning and evening on two consecutive days if kept upright in the refrigerator, Dr. Trattler said. Refrigeration is important to inhibit the growth of bacteria, and cold drops can be more comfortable to use. However, drops with preservatives may be more convenient to keep in a pocket or purse or to carry when traveling.

Natural tears and some artificial products have an oily component to help keep liquid in the eye, Dr. Trattler said. Patients should be encouraged to select drops that meet their needs in various settings. Drops needed when working outside will be different from those needed when viewing a computer screen indoors.

If inflammation is present, patients can try cyclosporine ophthalmic emulsion (Restasis). Ap-

proved by the Food and Drug Administration in 2003, Restasis increases tear production and has anti-inflammatory properties, making it a good choice for patients with occluded tear ducts, noted Ashley Behrens, M.D., of Johns Hopkins University in Baltimore.

However, patients should be cautioned when using Restasis, since the effects of overuse of the drug are unknown. In the study that served as the basis for the FDA approval, the drops were used twice daily, but "using them more frequently won't necessarily add benefits," Dr. Behrens said.

Both ophthalmologists, neither of whom reported a financial interest in any eye care product, recommended that Sjögren's patients use over-the-counter lubricant eye gels for periods of sleep. Warm compresses can provide relief, and specialized moisturizing goggles are available. In addition, patients should be advised to turn off ceiling fans at night.

In treatment-resistant cases, Sjögren's patients can have their tear ducts blocked with punctal plugs. The ducts drain the tears into the nose, and plugging them helps retain fluid in the eye. Cauterization of the tear ducts has the same result, but while plugs can be removed, cauterization permanently closes the ducts.

For patients who develop blepharitis, inflammation of the eyelids, Dr. Trattler recommends baby shampoo and warm compresses. The inflammation occurs when the oil that is a component of normal tears crystallizes on the lids, he explained. ■

## Skin Care: Limit Washing, Study Product Labels

FORT LAUDERDALE, FLA. — Skin manifestations of Sjögren's syndrome may not be unique to the disease, but they require specific interventions and close monitoring, Darren L. Casey, M.D., said at a meeting sponsored by the Sjögren's Syndrome Foundation.

Sjögren's syndrome affects the body's moisture-producing glands, resulting in chronically dry, itchy, and scaly skin.

Remind patients that they should try to maintain a protective barrier on the skin. "When we wash too aggressively, we get rid of that barrier," said Dr. Casey, a dermatologist in private practice in Atlanta.

Dr. Casey suggests that patients limit washing to 20 minutes and recommends using Cetaphil, Dove, or Oil of Olay gentle skin cleansers.

Use of emollients such as Aquaphor immediately after a bath or shower can help retain moisture if lotions and creams do not provide relief. ■

For patients who don't respond to over-the-counter products, prescription humectants such as Carmol 20 and Carmol 40 may help.

He also advises patients to use humidifiers and fragrance-free laundry detergents to reduce irritation of dry skin.

In addition to dry skin, Sjögren's patients may develop red or purple palpable spots related to small vessel disease.

Urticarial vasculitis, which affects some patients, is characterized by hives that last hours to days. When these lesions are biopsied, they show inflammation in the blood vessels.

Medium-sized vessel disease typically manifests as nodules on the hands and feet or as livedo reticularis, characterized by a whorl pattern of pigmentation brought on by constricted blood flow. Medium-sized vessel disease should be a red flag to investigate central nervous system or severe organ involvement. ■