

# CE429 | 1.0 hr

## The Desert of Sjögren's Syndrome

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### Course Objectives

The goal of this program is to provide nurses with information about the incidence, etiology, identification, and treatment of Sjögren's syndrome. After studying the information presented here, you will be able to —

- Discuss four extraglandular manifestations of Sjögren's syndrome.
- Describe four interventions for relief of dry eyes.
- Identify two medications used to treat dry mouth.

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Imagine having to give up outdoor activities that you love — from gardening to cross-country skiing — because your eyes are too dry. Imagine having to sip water all day to keep your mouth moist enough to speak and chew comfortably. Imagine having fatigue severe enough to limit your activities on the job — or even make it difficult for you to take care of your children. These are just some of the problems that people with Sjögren's syndrome can face.

Sjögren's is considered the second most common autoimmune disorder in the United States, second only to rheumatoid arthritis.<sup>1</sup> It is a chronic inflammatory condition of the exocrine glands (those with external secretions) that diminishes lacrimal and salivary gland secretions.<sup>2</sup>

Sjögren's syndrome affects mostly women (9:1) in their 40s and 50s.<sup>3</sup> It is named after Swedish physician Henrik Sjögren, who in 1933 noticed that many patients with arthritis also had dry mouth and dry eyes. Sjögren's affects about 4 million Americans<sup>4</sup> and is underreported.

Patients with Sjögren's typically take several years to be diagnosed, often after being shunted from practitioner to practitioner. Nurses can help speed this process by learning to recognize manifestations of Sjögren's and educating other providers as well as patients.

### Parched eyes and mouth

The hallmark signs of Sjögren's are the dry eye condition keratoconjunctivitis sicca (KCS) and the dry mouth condition xerostomia.<sup>2</sup> Sjögren's is also a systemic condition, and organs other than the eyes and mouth are affected. Some of the extraglandular symptoms include arthritis; Raynaud's syndrome; pulmonary involvement, such as pleural effusions and dyspnea; hepatobiliary features, such as primary biliary cirrhosis; renal involvement; and vasculitis with skin changes. In addition, patients with Sjögren's have a greater than normal risk of developing non-Hodgkin's lymphoma.<sup>3</sup>

Sjögren's syndrome can be primary, without concomitant disease, or secondary, with an associated condition, such as rheumatoid arthritis, scleroderma, multiple sclerosis, or systemic lupus erythematosus. The prevalence of primary and secondary Sjögren's syndrome is about equal.<sup>5</sup>

Many etiological factors are posited as causes of Sjögren's syndrome. Possibilities include viral, hormonal, genetic, and environmental factors.<sup>4</sup> The actual triggering factor is unknown although most researchers indicate that autoimmune conditions, such as Sjögren's syndrome, involve an interaction of specific genes and environmental agents.<sup>5</sup> What is known is that lymphocytes attack the moisture-producing glands, and programmed cell death of T lymphocytes occurs.<sup>5</sup> Next, salivary acinar (secretory) epithelial cells malfunction, causing the abnormal release of proteins that regulate immunological aspects of cell growth (tissue cytokines).<sup>5</sup> Research shows that a possible genetic predisposition for Sjögren's exists and that sex hormones may be

involved since Sjögren's occurs more often in women than in men.<sup>6</sup>

## Why dry?

It's important to understand the pathophysiology of the dry eyes and dry mouth found in Sjögren's. KCS is caused by inflammatory changes to the lacrimal glands and the ocular epithelium and by aqueous deficiency in the tear film of the eyes.<sup>1</sup> The dysfunctional lacrimal glands cannot keep up with loss from evaporation, leading to decreased lubrication and decreased tears. This also means decreased antimicrobial activity.<sup>1</sup> In the mouth, inflammation of the salivary glands may cause pain and swelling and can lead to a reduction in the production of saliva, depriving the oral cavity of saliva's antimicrobial properties.

It's upsetting to patients that the precise cause of Sjögren's is unknown; that is compounded by the lack of a cure. What starts out as mildly annoying dry eyes and mouth can gradually restrict a person's life as the syndrome progresses. Although remission can occur, most patients experience a steady diminution in their normal life functions, along with worsening symptoms. Family and friends often do not understand the severe dryness that their loved ones are experiencing and the extent of the disability caused by fatigue and myalgia. Sjögren's can severely reduce patients' quality of life, making early diagnosis crucial to minimize the impact.<sup>3</sup>

## A long wait

Sjögren's syndrome is not always easy to diagnose, and many patients wait years to learn their diagnosis. This long wait is due to the complicated array of symptoms, as well as healthcare professionals' lack of awareness. Classification criteria for syndromes are helpful to standardize diagnoses; the American-European Consensus Group has developed classification criteria for Sjögren's.<sup>7</sup>

Diagnostic tests: Diagnostic tests include those for the eyes, the mouth, and systemic manifestations. A [Schirmer's test](#) evaluates the amount of tear secretions. A paper strip is placed into the lateral third of the lower lid of each eye for five minutes. The amount of moisture on the paper measured, with less than 5 mm being definitive for dry eye. Other tests include the rose bengal or fluorescein dye tests, which use a stain and a slit lamp to observe the surface of the eye. Diagnostic tests for xerostomia include salivary flow rate measurement (sialometry), X-ray exam of the salivary duct system (sialography), and lip gland biopsy. Serology is used to identify the presence of specific antibodies (to anti-Ro and anti-La).<sup>3</sup> Some patients have an elevated sedimentation rate and low hemoglobin and hematocrit.<sup>3</sup> Many patients are positive for antinuclear antibodies.

Glandular manifestations: The course of Sjögren's varies greatly from patient to patient. Each person has a unique presentation, with varying degrees of dryness in the eyes and mouth and other signs and symptoms. But about 85% of patients present with sicca (dry) symptoms affecting the glands.<sup>1</sup>

Many uncomfortable ocular manifestations exist. Patients may complain of a dry, sticky, gritty, or burning feeling in their eyes. The watery component of tears is lacking, leading to thick mucous strands in the eyes.<sup>3</sup> Some patients complain of blurry vision and use eye drops frequently. The mouth is likewise affected: The patient may mention dry mouth, and on oral exam, the healthcare provider may notice a dry, sticky, furrowed tongue and a lack of saliva pooling under the tongue.

Patients may further complain of salivary gland enlargement, severe lip cracking (angular cheilitis), ulceration or burning of the tongue (stomatopyrosis), a sudden increase of dental caries, halitosis, dysphagia, altered taste, oral discomfort, and difficulty wearing dentures.<sup>3</sup> Eating a cracker or licking an envelope is impossible — even the thought of doing so is distasteful. Chronic candidiasis often accompanies dry mouth.

Women with Sjögren's may experience a dry vagina as a result of infiltration of vaginal tissue with chronic inflammatory cells. This may lead to [vulvodinia](#) (chronic vulvar itching, burning, and pain that causes physical and psychological distress) and dyspareunia (painful sexual intercourse), even in premenopausal women. Other dry areas are the ear canal and the nose, leading to itching and nasal sores.

Extraglandular manifestations: Extraglandular problems are common in Sjögren's. Disabling fatigue is a major complaint and contributes to poor quality of life. Fatigue may be accompanied by lethargy, depression, cognitive

dysfunction, and irritability, possibly aggravated by poor sleep, nocturia, and muscle aches. Skin problems may appear as dryness and rashes, such as vasculitis and purpura. Pulmonary manifestations include dry cough, interstitial pneumonitis, pulmonary hypertension, and pulmonary embolism.<sup>8</sup> Sinus infections are common as a result of thickened secretions. Patients may have renal manifestations, such as interstitial nephritis (inflammation of the kidney) and glomerulonephritis (inflammation of the glomeruli).<sup>8</sup> One of the most serious systemic manifestations is the patient's higher-than-normal risk of developing non-Hodgkin's lymphoma, a cancer of the lymph nodes.<sup>3</sup>

Any patient with known autoimmune disease is at risk for developing other autoimmune conditions. Accompanying autoimmune disorders found in Sjögren's patients include [celiac disease](#), primary biliary cirrhosis, chronic active autoimmune hepatitis, myasthenia gravis, pernicious anemia, multiple sclerosis, Addison's disease, Graves' disease, and Hashimoto's thyroiditis.<sup>9</sup>

## Seeking moisture

Treatment for Sjögren's involves the management of dry eyes, dry mouth, and systemic manifestations:

**Dry eye management:** Treatment for dry eye includes several modalities. In 2004, an international task force issued evidence-based guidelines for patient management.<sup>10</sup> (See illustration.) Most patients with dry eye will receive symptomatic benefit initially from artificial tears (preserved or unpreserved) that augment tear films, lacrimal inserts (a small pellet that dissolves and spreads a thin film over the eye), and OTC lubricating gels and ointments.<sup>5</sup> Good lid hygiene involves cleaning the eyelids carefully with baby shampoo, using hot compresses over the eyes, and avoiding excessive eye makeup, especially on the eyelid.<sup>5</sup> Punctal occlusion — either temporary (plugs) or permanent (cautery) — and special moisture goggles promote retention of tear fluids on the ocular surface.<sup>5</sup>

Pharmacological treatment involves the topical use of doxycycline and tetracycline, antibiotics with antiinflammatory properties,<sup>10</sup> and cyclosporine A (Restasis) ophthalmic drops.<sup>10</sup>

A mainstay of treatment for both dry eyes and mouth is the use of secretagogues, medications that enhance secretion by stimulating certain receptors of the exocrine glands. Two agents approved for use as secretagogues are pilocarpine (Salagen) and cevimeline (Evxac), which stimulate the acinar and ductal cells of the salivary and lacrimal glands.<sup>11</sup> Pilocarpine (5 mg tid) and cevimeline (15mg to 30 mg tid) work by preventing programmed cell death. They also diminish the damage caused by proinflammatory cytokines while optimizing the function of residual glandular cells in patients with Sjögren's.<sup>11</sup> Healthcare providers should use caution in administering these medications to patients with asthma, narrow-angle glaucoma, acute iritis, severe cardiovascular disease, biliary disease, nephrolithiasis, diarrhea, or ulcer disease.<sup>11</sup> Adverse effects of pilocarpine are primarily excessive sweating, nausea, rhinitis, and diarrhea related to its secretory-stimulating properties.<sup>11</sup> Sweating as an adverse effect is somewhat lessened with the use of cevimeline.

**Dry mouth management:** Many treatments exist for dry mouth, for example, artificial saliva, which is similar to artificial tears. But since the effectiveness of artificial saliva is short-lived, it needs to be used only by those who have no residual salivary function. Other products include the OTC oral moisturizing gel Oralbalance; the new OTC product Oasis, in the form of mouthwash and mouth spray; and the new prescription liquid and lozenge Numoisyn.<sup>11,12</sup>

Patients must be scrupulous about oral care. To avoid gingivitis and dental caries, patients should brush and floss their teeth frequently, use fluoride mouth rinses and toothpastes, and have dental examinations and cleanings every two or three months. Keeping the lips moist with a petrolatum-based product is helpful. Patients also must take care to avoid oral candidiasis and treat it promptly if it occurs.

Sjögren's patients should avoid medications that have drying effects, such as diuretics, antihypertensives, antidepressants, antihistamines, and anticholinergics.<sup>11</sup> Patients should also avoid anything with external drying effects, such as excessive air conditioning, forced-air heat, and wind or a dry climate. Patients should increase their exposure to humidified air by using a mechanical humidifier in the bedroom, which will make it easier for them to sleep through the night. Drinking enough water and using gum or lozenges (sugarfree to prevent caries)

are additional moisturizing tips.

## Bodywide treatments

Many medications help treat the systemic effects of Sjögren's. Nonsteroidal antiinflammatory drugs may provide some relief from myalgias and the pain of swollen parotid glands. Hydroxychloroquine (Plaquenil) is somewhat helpful for immune hyperreactions (hypergamma-globulinemia and autoantibody levels) and can be used for myalgias and arthralgias.<sup>11</sup> Corticosteroids (Prednisone) can be used for their antiinflammatory properties in patients with painful joints, vasculitis, and renal tubular acidosis.<sup>11</sup>

Other helpful medications include antidepressants (not tricyclics, which cause dryness); hypnotics; and anxiolytics, especially for patients with depression, sleep problems, and anxiety related to Sjögren's.<sup>11</sup> Antacids, histamine H2 receptor blockers, and proton pump inhibitors can be used for gastroesophageal reflux. Secretagogues and guaifenesin (Robitussin) can help relieve dryness in the throat and trachea.<sup>11</sup> For dry vagina, patients may try the compound polycarbophil found in the OTC products Replens and Durex Sensilube, which release water to rehydrate epithelial cells and keep the vagina moist for longer periods.<sup>13</sup> Estrogen creams and oral estrogen may relieve vaginal dryness. Finally, immunomodulating agents, such as methotrexate (Rheumatrex), cyclophosphamide (Cytoxan), and azathioprine (Imuran), may be used in the most severe cases.<sup>5</sup>

Nonpharmacologic management: Many nonpharmacologic approaches are available for patients with Sjögren's. Humidification is helpful for dry eyes, mouth, and skin. Reducing the water temperature in showers and baths and using moisturizing agents for the skin can be beneficial. Vitamin E oil for the nose, ears, vagina, and even the tongue and lips is especially helpful at bedtime. Saline irrigations for the nose help reduce thick, crusty mucosal secretions. Patients should avoid alcohol and mouthwashes that contain alcohol, as well as mouth breathing.<sup>5</sup> While drinking adequate water is important, so is altering the diet to avoid overly dry, rough, spicy, and hot foods. Eating small, frequent meals may be easier for patients with Sjögren's. Exercise, interspersed with rest periods and adequate sleep, is crucial.

Female patients with Sjögren's may have to make changes in their sexual relationships. Premenopausal women may find it advantageous to choose certain times of the month when their natural lubrication is higher to have intercourse. Reducing the length of time of intercourse and even choosing nonpenetration may be necessary. Healthcare providers should reassure patients and their partners that Sjögren's-induced vaginal dryness is a physiological problem, not related to performance or response.<sup>13</sup>

Perioperative patients: Patients with Sjögren's who have surgery may have to make changes in their regular treatment regimen. Two weeks before surgery, they should discontinue any vitamin E oil they may be taking to avoid a possible anticoagulant effect.<sup>5</sup> Because they already have diminished salivary secretions, a prolonged NPO status and anticholinergic drugs (atropine, glycopyrrolate [Robinul]) and antihistamines (promethazine [Phenergan], diphenhydramine [Benadryl]) should be avoided.<sup>5</sup> Anesthetic gases are drying, so local or regional anesthesia is preferred.<sup>5</sup> Patients will need ointment in their eyes and lubrication on their lips pre- and postoperatively. The OR temperature may need to be raised to avoid a flare of Raynaud's phenomenon, and positioning is important for patients with joint and muscle pain.<sup>5</sup>

After surgery, patients must have all their medications available to use for lubrication as needed. Humidified oxygen should be delivered along with ice chips or liquids as soon as possible. Give oral medications with enough water for comfort; healthcare providers may need to give medications in an alternative form if patients have difficulty swallowing.<sup>5</sup>

Patients can benefit from referral to a support group and membership in the [Sjögren's Syndrome Foundation](#). The foundation publishes a monthly newsletter, The Moisture Seekers, for patients. For healthcare providers there is Sjögren's Quarterly and an allied health professionals' group, which includes nurses, that meets every other month via conference call. To find out more, [contact the foundation](#).

## Offering hope

Nurses are in an excellent position to help Sjögren's patients in primary, acute, and long-term care settings. But

helping patients with physical symptoms and treatments is not enough. Nurses also need to support patients emotionally. In a qualitative research study involving interviews with 10 women with Sjögren's, this author found that the women had many fears about lifestyle changes and possible future incapacitation.<sup>14</sup> They wanted to learn all they could about their condition, educate others, and have their voices heard. (In the process of being diagnosed, many of these women felt they were not heard.<sup>14</sup> Nurses can be the listening ear, the support that people living with a chronic illness syndrome need.

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