**SJÖGREN’S SYNDROME**
(also known as “Sicca Syndrome”, “SS”, and “Gougerat-Sjögren Syndrome”)

**Is the initiation of non-invasive dental hygiene procedures* contra-indicated?**  No

- Is medical consult advised? No (assuming patient/client is already under medical care for SS and there is no suspicion of malignancy)

**Is the initiation of invasive dental hygiene procedures contra-indicated?**  Possibly, but not typically

- Is medical consult advised? …………………………………. See above.
- Is medical clearance required? …………………………………. Possibly, if patient/client is being treated with medications associated with immunosuppression +/- increased risk of infection (e.g., corticosteroids [e.g., prednisone], methotrexate, cyclosporine, azathioprine, mycophenolate, cyclophosphamide, and rituximab)
- Is antibiotic prophylaxis required? …………………………… No
- Is postponing treatment advised? …………………………… Possibly but not typically (depends on medical clearance for patients/clients on medications associated with immunosuppression)

**Oral management implications**

- While most patients/clients with dry mouth do not have SS, the dental hygienist should be alert to the possibility of undiagnosed Sjögren’s syndrome, particularly if the patient/client also has dry eyes, dryness of other mucosal surfaces, history of salivary gland swelling, and numbness or altered sensation of the head and neck. Rampant dental caries, particularly in middle-aged women, is another possible indicator of SS. Clinical suspicion should prompt medical referral for investigation and definitive diagnosis.
- Factors that contribute to hyposalivation should be addressed, including smoking, poor diabetes control, and many drugs. The dental hygienist can play a role in this by encouraging an appropriate medical consult. In addition, avoidance of diuretics such as caffeine and alcohol should be encouraged.
- Hyposalivation management includes frequent sipping of water, chewing sugar-free gum, sucking sugar-free candy, use of oral lubricants and moisturizing mouth rinses, and saliva replacement therapy. Lemon juice in water can stimulate salivary flow. Further information on management can be found in the CDHO’s Xerostomia Fact Sheet. Medical or dental referral may be considered for possible pharmacologic (e.g., pilocarpine or cevimeline) stimulation of the salivary glands; appropriate sialagogue drug therapy may be more effective than carboxymethylcellulose- or hydroxymethylcellulose-based saliva substitutes that are often too viscous or insufficiently viscous for patients/clients with SS.
- A nasal saline spray can help moisturize and clear nasal passages so the patient/client can breathe more freely through the nose and decrease mouth breathing. Use of a humidifier at night may also make the xerostomia more tolerable.
- The risks of dental caries and enamel erosion are elevated, and thus daily fluoride applications and brushing with casein phosphopeptide-amorphous calcium and phosphate products should be considered. Use of minimally abrasive fluoridated dentifrices and irrigation devices should be encouraged. The interval between dental hygiene recall appointments should be sufficiently short to ensure early detection and treatment of root caries.
- Persons with Sjögren’s syndrome, as with persons with other causes of xerostomia, are at increased risk of oral candidiasis. The dental hygienist should be alert to signs and symptoms of oral candidiasis and ensure the patient/client is referred for medical/dental treatment, if indicated.

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Disease/Medical Condition

SJÖGREN’S SYNDROME

(also known as “Sicca Syndrome”, “SS”, and “Gougerat-Sjögren Syndrome”)

Oral management implications  (cont’d)

- Dental hygienists should be alert to swelling of the salivary glands, which could be a manifestation of lymphoma, particularly in patients with progression of the primary form of Sjögren’s syndrome. SS patients/clients are also at increased risk of lymphoma development in the oral cavity itself. Prompt medical referral is warranted if lymphoma is suspected.
- SS-induced thrombocytopenia that results in oral bruising or purpura may complicate oral health care.
- Occasionally, patients/clients may experience intraoral or facial paresthesia or anesthesia, which should prompt medical referral.
- Due to difficulty tasting and swallowing certain foods, some patients/clients may have inadequate dietary intake of certain nutrients. Dental hygienists should be alert to this possibility and consider medical/dietary referral where appropriate.

Oral manifestations

- Dry mouth (in addition to dry eyes) is a characteristic manifestation. The salivary glands (as well as the lacrimal glands) are usually affected first, resulting in decreased production of saliva (and tears).
- Oral signs and symptoms are the first manifestation for more than half of SS patients/clients, with most resulting from hyposalivation. “Cotton mouth” is a common descriptor used by patients/clients of their oral symptoms; lack of saliva causes the mouth to feel sticky.
- Saliva may be frothy rather than liquid in consistency. Submandibular and parotid expression of saliva may be reduced or absent. Intraoral examination may reveal absence of salivary pooling in the floor of the mouth.
- The tongue may be sore, erythematous, dry, and fissured or be coated and brown or black (i.e., black hairy tongue). There is generalized atrophy of both filiform and fungiform papillae. Lips are cracked and dry.
- Hyposalivation can cause difficulty with speaking, eating, chewing, swallowing, and retaining dentures.
- Oral mucosa, which may be normal or erythematous in appearance, may be dry and tend to stick to the dental mirror or gloved finger. Secondary candidiasis is common, as are angular cheilitis and denture sores.
- Halitosis, dysgeusia (altered taste), and new, recurrent or atypical patterns of dental caries (especially cervical caries at the gum line) may occur. Periodontitis and gingivitis are common. Patients/clients tend to have higher decayed/missing/filled (DMF) teeth scores than persons without SS.
- Up to half of patients/clients with SS develop diffuse, firm enlargement of the parotid salivary glands during the course of their disease. The swelling is usually bilateral and can be nonpainful or slightly tender; it may be persistent or intermittent. The likelihood of salivary gland enlargement tends to correlate with severity of the disease. Reduced salivary flow also increases the risk of retrograde bacterial sialadenitis, which may present with fever, acute swelling, pain, regional lymphadenopathy, and pus discharge.
- Trigeminal neuropathy and other neuropathies can develop in Primary Sjögren’s Syndrome.
- Systemic involvement can include SS-induced thrombocytopenia, which manifests as oral bruising or purpura.

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Disease/Medical Condition

SJÖGREN’S SYNDROME

(also known as “Sicca Syndrome”, “SS”, and “Gougerat-Sjögren Syndrome”)

Oral manifestations (cont’d)

- Persons with the primary form of Sjögren’s syndrome (PSS) are at greatly elevated risk of developing B-cell lymphoma of the salivary glands, which typically manifests as swelling.
- Persons with the secondary form of Sjögren’s syndrome (SSS) also may have oral manifestations related to their primary autoimmune disease, such as systemic lupus erythematosus or scleroderma.
- Certain disease-modifying anti-rheumatic drugs (DMARDs) may be used to treat severe SS that progresses to involve the skin, lungs, kidneys, and/or other organs. These DMARDs, such as cyclosporine, have side effects, which include bleeding, oral ulcerations, stomatitis, and tender or swollen gums. Methotrexate can cause mouth sores, in addition to sore throat. Hydroxychloroquine may lead to reversible pigmentary changes of the mucous membranes.

Related signs and symptoms

- Sjögren’s (SHOW-grins) syndrome (SS) is a chronic, systemic autoimmune disorder with prevalence in the general population of about 1%. Nearly 90% of cases are first diagnosed in middle-aged women (usually after age 45), with children rarely being affected. Treatment focuses on relieving symptoms, which may subside or worsen with time. There is no known cure.
- In addition to dry mouth (xerostomia), dry eyes (xerophthalmia) are a characteristic manifestation. The mucous membranes and moisture secreting glands of the eyes (and mouth) are usually affected first, resulting in decreased production of tears (and saliva). Patients/clients with SS complain of eye symptoms such as grittiness, blurred vision, itchiness, and discomfort; conjunctival redness may be a visible sign. Adverse environmental factors exacerbate the ocular symptoms, which tend to become worse later in the day. Corneal and conjunctival epithelial changes may occur.
- Primary Sjögren’s syndrome (PSS) is a systemic autoimmune disorder marked by inflamed exocrine glands (especially salivary and lacrimal) but without the presence of another autoimmune connective tissue disease. Secondary Sjögren’s syndrome (SSS) has a similar presentation (i.e., dry mouth and eyes), but is associated with other autoimmune connective tissue diseases, particularly rheumatoid arthritis and systemic lupus erythematosus. About 50% of patient/clients with SS have another autoimmune disease. Associated conditions also include primary biliary cirrhosis, fibromyalgia, mixed connective tissue disease, polymyositis, scleroderma, and Raynaud’s syndrome.
- In addition to the orofacial symptoms, patients/clients may have general malaise, chronic fatigue, dry skin, myalgia, arthralgia, vulvar and vaginal dryness, lymphadenopathy, hematologic problems such as anemia and macroglobulinemia, gastrointestinal problems (especially gastro-esophageal acid reflux), peripheral neuropathy (especially of the sensory nerves, resulting in tingling and burning sensations in the hands and feet), interstitial nephritis, interstitial lung fibrosis, vasculitis, Raynaud phenomenon (i.e., extreme sensitivity to cold in the fingers and toes resulting in blanching), and pancreas and heart involvement (e.g., pulmonary hypertension). Inflammation may also contribute to pneumonia and bronchitis, as well as autoimmune hepatitis and primary biliary cirrhosis.
- PSS patients/clients are at greatly elevated risk (44 times normal) for developing malignant lymphomas, particularly non-Hodgkin’s lymphoma. Whereas most PSS-associated lymphomas arise from the salivary glands, overall most lymphomas are of extranodal origin in persons with SS and arise in the mucosa-associated lymphoid tissue (MALT).
Disease/Medical Condition

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References and sources of more detailed information

- Mayo Clinic  http://www.mayoclinic.org/diseases-conditions/sjogrens-syndrome
- American College of Rheumatology  http://www.rheumatology.org/Practice/Clinical/Patients/Diseases_And_Conditions/Sjogrens_Syndrome/
- Sjögren’s Society of Canada  http://sjogrenscanada.org/answers/
- National Health Service (U.K.)  http://www.nhs.uk/Conditions/Sjogrens-syndrome/Pages/Treatment.aspx

* Includes oral hygiene instruction, fitting a mouth guard, taking an impression, etc.

** Ontario Regulation 501/07 made under the Dental Hygiene Act, 1991. Invasive dental hygiene procedures are scaling teeth and root planing, including curetting surrounding tissue.

Date: April 15, 2014