SCLERODERMA

(includes “systemic scleroderma” [also known as “systemic scleroderma” and “systemic sclerosis” or “PSS”; includes “limited cutaneous”, “diffuse cutaneous”, “systemic scleroderma sine”, and “environmentally-induced” types] and “localized cutaneous scleroderma” [also known as “localized scleroderma”; includes “morphea” and “linear” types])

Note: This fact sheet focuses on systemic scleroderma unless otherwise indicated.

Is the initiation of non-invasive dental hygiene procedures* contra-indicated? No (assuming stem cell transplantation is not involved)

- Is medical consult advised? No (assuming patient/client is already under medical care for scleroderma, systemic manifestations are well managed, and there are no stem cell transplantation considerations).

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

- Is medical consult¹ advised? Possibly (depends on severity and level of control of the disease).
- Is medical clearance required? Possibly, if patient/client is being treated with medications associated with immunosuppression +/- increased risk of infection (e.g., corticosteroids [such as prednisone], cyclophosphamide, cyclosporine, mycophenolate mofetil, methotrexate, rituximab, and antithymocyte globulin). Medical clearance may also be required if the patient/client is taking D-penicillamine, which can cause thrombocytopenia (low platelet count). In severe cases of scleroderma, stem cell transplantation may be a therapy, in which case medical clearance is required.
- Is antibiotic prophylaxis required? No, not typically (although extended use of corticosteroids or cytotoxic drugs — particularly in the presence of leukopenia [low white blood cell count] — may warrant consideration of antibiotic prophylaxis).
- Is postponing treatment advised? Possibly, but not typically (depends on severity and level of control of the disease, as well as medical clearance for patients/clients with thrombocytopenia or on medications associated with immunosuppression).

Oral management implications

- The dental hygienist may be the first person to note the manifestations of scleroderma. Early recognition, medical referral, and treatment are important to reduce the disease’s destructive impact on oral tissues and other body systems. The most critical period for systemic scleroderma is usually within the first few years of onset.
- Patients/clients with scleroderma are at increased risk of oral disease, and early interventions are important to prevent systemic complications caused by oral infection.
- Dental hygiene appointments may need to be kept short, because as scleroderma progresses, an increasingly debilitated patient/client may not tolerate much time in the dental chair (particularly oral stress due to limited facial movement). Conversely, treatment interventions may take more time and be more complicated due to perioral fibrosis, which limits opening of the mouth.
- Given that many patients/clients with scleroderma struggle to keep their mouths open long enough to receive dental hygiene care, a mild analgesic taken an hour before the appointment may make them more comfortable.

¹ Persons with scleroderma are typically followed by a dermatologist (skin specialist) and/or rheumatologist (musculoskeletal and rheumatic disease specialist). Depending on disease complications and progression, a respirologist, orthopedic surgeon, gastroenterologist, and/or cardiologist may also be involved in care, in addition to the family physician.
Oral management implications (cont’d)

- A dental assistant may be needed to retract the tongue and assist with suction. A mouth prop will also aid the dental hygienist in gaining access by assisting the patient/client in keeping their mouth open.

- The semi-supine chair position is indicated where there are gastrointestinal problems, such as tendency for gastroesophageal reflux.

- Humidification of the treatment area may be beneficial, because many patients/clients with systemic scleroderma experience severe dryness. A room humidifier will help keep tissues hydrated, thereby easing skin tension and improving movement of tight skin.

- Supplemental oxygen may be required in patients/clients with severe restrictive lung disease.

- Because most patients/clients with PSS (and many with localized scleroderma) also have Raynaud’s phenomenon (which is exacerbated by cold and anxiety), the dental hygienist should consider increasing room temperature and keeping a blanket and gloves (or hand warmer) handy. Stress reduction for the patient/client may be achieved by soothing music, effective pain control, and facilitative guidance during the appointment. In some cases, anti-anxiety medication may be indicated.

- Ultrasonic scaling and air polishing may be inappropriate if there is significant fibrosis of the hard and soft palates.

- Injections by the dentist may be difficult due to induration of oral tissues, with associated discomfort for the patient/client. Furthermore, local anesthetics without epinephrine should typically be used, because epinephrine can exacerbate microangiopathy found in patients/clients with systemic scleroderma.

- Radiographs may be difficult to take in the presence of microstomia. Consequently, size one or zero film or sensors should be used, with only a few radiographs being exposed per visit. Panoramic radiographs should be taken periodically to screen for oral bone resorption.

- Immobility and rigidity of the patient/client’s skin affects oral facial structures and the hands, which can interfere with oral self-care. A pediatric toothbrush may improve access, although lip and gingival retraction on the buccal side of the teeth can make brushing even with a pediatric brush difficult. A tuft brush may be better.

- In order to increase mouth opening over time, the patient/client can be encouraged to lubricate the mouth, cross arms at chest height with palms facing down, and use the thumbs to stretch the corners of the mouth for several minutes. Mouth stretching using tongue depressors may improve mandibular range of motion. As well, isometric exercises may improve mouth opening and maintain mobility of the face and neck. Physiotherapist involvement is often beneficial.

- The patient/client’s oral hygiene education and regimen should take into account loss of manual dexterity2 and strength secondary to fibrosis of the fingers, which compromises brushing and flossing abilities. In such cases, oral hygiene devices may need to be modified (e.g., large flat-handled toothbrushes) or replaced with power instruments (e.g., electric toothbrushes and flossing devices). Finger cots may improve oral self-care capability, and dental tape may be easier to grasp than Teflon-coated floss (which is too slippery for some patients/clients). Stretching exercises for the hands can also be helpful, and some patients/clients may benefit from physical and/or occupational rehabilitation.

- Frequent professional examinations, cleanings, and periodontal evaluations are indicated. Patients/clients with systemic scleroderma often consume a soft, high carbohydrate diet in light of their dysphagia, which predisposes them to caries. As well, fatigue, pain, and depression may affect patients/clients’ ability to perform oral self-care.

- Patients/clients taking calcium channel blockers need to be monitored for gingival overgrowth.

- In addition to nutritional counselling, caries risk reduction can be achieved via fluorides, remineralization therapy, and xylitol.

- Xerostomia should be addressed by salivary replacement therapies and associated measures.

- Antimicrobial mouth rinses may be indicated in some patients/clients.

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2 Loss of manual dexterity is a particular problem in patients/clients with CREST syndrome (see “Oral Manifestations” below), because the deposition of calcium salts in the tissues leads to thickening and tightening of the skin in the extremities.
Oral manifestations

- The head and neck region is involved in more than 70% of patients/clients with systemic scleroderma.
- Periodontal disease and caries risks are elevated in patients/clients with scleroderma.
- Depending on the extent of the disease, the patient/client may exhibit microstomia (constriction of the mouth) and an inability to close the lips (lip retraction). These manifestations are due to perioral fibrosis, and fibrotic tissue may be seen as a tan band around midline-inner part of the lip. As well, radial furrowing around the lips, hyperpigmentation, and loss of skin folds around the mouth are often seen. Anterior open bite is an associated finding.
- Fibrosis of the tongue and of the hard and soft palates may occur. Rigid, hypomobile tongue contributes to difficulties with swallowing and speech.
- Upward retraction and shortening of the uvula may occur, as may atrophy and blanching of the oral mucosa.
- Loss of gingival mucosa attachment and generalized gingival recession are seen in CREST syndrome.\(^3\)
- Xerostomia results from decreases in quantity and quality of saliva, which are caused by fibrosis of the salivary glands. As a result, risk of cervical caries is increased and dry mucous membranes are common. Speech, swallowing, and denture retention are also adversely impacted. As well, oral candidiasis may accompany xerostomia.
- Intermittent salivary gland enlargement sometimes occurs in PSS.
- Asymptomatic resorption of mandibular bone is common in PSS. Such resorption, however, increases the risk of mandibular fracture, which is seen late in the disease process.
- Resorption of teeth occasionally occurs.
- Widened periodontal ligament (PDL) spaces (seen in periapical radiographs) are characteristic manifestations of systemic scleroderma. Other radiographic findings include osseous destruction/resorption of the TMJ, mandibular angle (“blunting”), condyle, or coronoid process.
- PDL space enlargement usually affects all teeth, and the extent is related to disease severity. Enlargement tends to be more pronounced on posterior teeth.
- Temporomandibular joint (TMJ) problems may arise due to tissue constriction. Dense perioral fibrosis causes trismus.
- Trigeminal neuropathy may result from excess collagen accumulation in the perineurium and reduced vascularity of the nerve itself. Intense flashes of pain can be triggered by contact with the cheek or by brushing teeth, eating, drinking, or talking.
- Perimylyolysis\(^4\) can be seen in scleroderma-associated GERD.
- Gingival hyperplasia may result from calcium channel blocker\(^5\) drug treatment of scleroderma and Raynaud’s phenomenon.

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3 CREST stands for calcinosis, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia.
4 Decalcification of the teeth due to gastric acid exposure
5 Calcium channel blockers are used to relax blood vessels and hence improve blood circulation. They include nifedipine, amlodipine, felodipine, and isradipine.
Disease/Medical Condition

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Oral manifestations (cont’d)

- Dysgeusia (altered or decreased taste) and lip swelling may be caused by D-penicillamine, a drug with anti-fibrotic properties sometimes used to treat scleroderma. D-penicillamine can also predispose the patient/client to oral bleeding.
- The use of corticosteroids (such as prednisone) in the treatment of scleroderma can lead to oral candidiasis (yeast infection) and other oral infections as a result of immunosuppression.
- Some disease-modifying drugs, 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. Rituximab can cause painful ulcerations in the mouth and on the lips.

Related signs and symptoms

- Scleroderma is a chronic disorder characterized by excessive accumulation and sclerosis (hardening and thickening) of the connective tissues. In its systemic form, there is also vascular dysfunction and dysregulation of the immune system.
- Scleroderma affects about 30,000 persons in Canada, with about 1/3 of cases being progressive systemic sclerosis (noticeable onset occurring most often in adults aged 30 to 55 years) and the remaining 2/3 being localized cutaneous forms (the linear type occurring most often in children and adolescents and the more common morphea type in women aged 20 to 40 years). PSS involves both skin and organs, and some persons with PSS also exhibit CREST syndrome.
- Similar to most autoimmune disorders, females are affected more often than males (4:1 in PSS). Other autoimmune disorders (including Sjögren’s syndrome, lupus erythematosus, and rheumatoid arthritis) may exist concurrently with systemic scleroderma, resulting in overlap syndromes.
- Environmental triggers (such as vinyl chloride, epoxy resins, paint solvents, and pesticides) or drugs (e.g., bleomycin) contribute to onset of systemic scleroderma in a minority of cases.
- Currently there is no definitive treatment that controls the underlying problem – the overproduction of collagen - common to all forms of scleroderma. Corticosteroids and immunosuppressive drugs benefit some persons.
- In localized cutaneous scleroderma, only the skin is involved, and onset is usually gradual. The morphea type is typified by firm, hardened, and discoloured (often purple-brown) oval patches of skin, usually with hair loss in affected areas. Sometimes there is also muscle and joint pain. While disfiguring, morphea tends to be otherwise relatively benign. By contrast, the linear type tends to have more severe sequelae. It manifests as long, narrow areas of thickened and discoloured skin, most often affecting the lower limbs and torso, and, less frequently, the hands, arms, and face. In severe cases, contracture deformities occur, which limit movement of affected areas.
- Systemic scleroderma is potentially life threatening, and sometimes rapidly progressive. In PSS, the disease can affect virtually any organ (in addition to skin and blood vessels) and may progress to affect many organ systems. The skin is typically affected first, although sometimes joint involvement may be the initial sign. Furthermore, Raynaud’s phenomenon sometimes precedes other manifestations of PSS, including skin tightening. As fibrosis of organs progresses, signs of organ failure (e.g., pulmonary, cardiac, and/or renal) begin to appear.
- Occasionally, scleroderma manifests in the internal organs without any cutaneous manifestations (“systemic scleroderma sine scleroderma”).

6 Raynaud’s phenomenon is a peripheral vascular condition characterized by spasm of the blood vessels. The fingers and toes are most often affected. Although it may be idiopathic, it is often associated with scleroderma and other autoimmune diseases such as lupus. Most patients/clients with systemic scleroderma have Raynaud’s phenomenon. Digital ischemia may be triggered by cold or emotional stress, and it is relieved by heat. Classically during an attack, the patient/client’s fingers are cold, cyanotic, painful, and somewhat rigid.

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Disease/Medical Condition

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Related signs and symptoms (cont’d)

- Cutaneous manifestations of PSS include pitting edema early in the disease process, followed by rigidity and tightness of the skin. Eventually, the skin becomes hard, smooth, and atrophic, often with telangiectasias\(^7\) and areas of hypopigmentation. Painful tightening and swelling of the skin occurs with resultant reduction in range of motion. The face becomes expressionless and resembles a mask, and the eyes may appear narrowed and the nose shrunken or pinched. In other cases, widened palpebral fissures may be associated with an inability to close the eyelids, exacerbating dryness of the eyes.

- Fibrosis of the fingers results in stiffness and atrophy of the skin over the digits, and the tightening of the skin’s connective tissue may cause the fingers to become fixed in a distorted position (“sclerodactyly” or claw-like deformity). Ischemia and ulceration of the fingertips may result from vascular compromise.

- Joint and muscle pain are experienced by most persons with diffuse systemic scleroderma. As well, severe skin dryness and itching are common, because the excess collagen in the dermis overwhelms the sweat and sebaceous glands.

- In CREST syndrome, esophageal dysmotility results from fibrosis and atrophy of smooth muscles. Patients/clients may exhibit hoarseness and wheezing, and as the disease progresses, gastroesophageal reflux disease (GERD) can occur, sometimes resulting in stricture formation.

- Malabsorption of nutrients, weight loss, and diarrhea or constipation may result from smooth muscle disruption in the intestine.

- Trigeminal neuralgia\(^8\) may occur.

- Dry eyes are common, particularly in PSS patients/clients who also have Sjögren’s syndrome.

- Erectile dysfunction, painful intercourse, or pregnancy complications may occur in persons with systemic scleroderma.

- In many patients/clients, systemic disease stabilizes after a time. However, continued progression can lead to death, with restrictive lung disease (and associated pulmonary hypertension) being the leading cause. Severe renal disease develops in up to 15% of persons with PSS, leading to acute renal failure (“scleroderma renal crisis”) as well as chronic kidney disease. Hypertension needs to be aggressively managed.

- Most patients/clients with localized scleroderma have a good prognosis and normal lifespan.

- Depression occurs in some patients/clients when they live with a severe, chronic disease.

References and sources of more detailed information

- RDH Magazine, Dentistry IQ Network


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7 Telangiectasias are vascular lesions of dilated small blood vessels, typically appearing as red to purplish pinpoint spots on the skin.

8 Trigeminal neuralgia, also called *tic douloureux*, is a chronic facial pain condition resulting from injury to the fifth cranial nerve.
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References and sources of more detailed information

- Scleroderma Canada [http://www.scleroderma.ca]
- Scleroderma Society of Ontario [http://www.sclerodermaontario.ca]
- Raynaud’s and Scleroderma Association (U.K.) [http://www.scleroderma.ca/pdf%20pages/RAS_Dental_Aspects_Scleroderma.pdf]
- Scleroderma & Raynaud’s UK [https://www.sruk.co.uk/scleroderma/scleroderma-and-your-body/oral-and-dental/]
- The Johns Hopkins Scleroderma Center [https://www.hopkinsscleroderma.org/patients/scleroderma-treatment-options/]

* 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: February 3, 2017