Disease/Medical Condition

PEMPHIGUS

(includes “pemphigus vulgaris” [“PV”], “pemphigus vegetans”, “pemphigus foliaceus”, “pemphigus erythematosus”, “paraneoplastic pemphigus”, and “drug-related pemphigus”)

Date of Publication: June 20, 2017

Unless otherwise indicated, this Fact Sheet focuses on pemphigus vulgaris, the most common type.

Is the initiation of non-invasive dental hygiene procedures* contra-indicated? No (assuming patient/client is already under medical care for pemphigus).

- Is medical consult advised?
  - Yes, if suspect oral lesions have not yet been diagnosed. Biopsy is necessary for a definitive diagnosis.
  - Yes, if not already done, all patients/clients diagnosed with oral pemphigus lesions should be referred to a dermatologist, as well as an ophthalmologist for assessment of ocular lesions.
  - Yes, if the disease is poorly controlled (as evidenced by severity and extent of mucous membrane and skin involvement).
  - Yes, if after treatment and remission by a specialist, new lesions are observed. The dental hygienist should ensure referral back to the physician for re-evaluation and further treatment.

Is the initiation of invasive dental hygiene procedures contra-indicated?** Possibly, depending on disease control and treatment regimen.

- Is medical consult advised? See above. Also, consultation may be appropriate for consideration of prophylactic prednisone prior to invasive procedures in order to reduce disease flares.
- Is medical clearance required? Possibly (e.g., if the disease is unstable and/or oral involvement is severe). Also, medical clearance may be required if patient/client is being treated with medications associated with immunosuppression plus/minus increased risk of infection (e.g., systemic corticosteroids [i.e., intravenous formulations or oral prednisone, methotrexate, azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil, chlorambucil and biologics [e.g., rituximab, infliximab, adalimumab, and etanercept]). Thrombocytopenia (low platelet count) is a side effect of many non-steroidal immunosuppressive drugs, and medical clearance may be required to rule out significant bleeding risk.
- 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 (depends on severity and level of control of the disease, as well as medical clearance for patients/clients on medications associated with immunosuppression and/or thrombocytopenia).

Oral management implications

- In up to 80% of cases of pemphigus vulgaris, the first signs of disease occur in the oral cavity (and in about ¼ of cases the oral mucosa is the only affected site). Typically, patients/clients will have multiple oral ulcers that persist for weeks to months. The mouth lesions often precede skin lesions by periods up to a year, and oral health professionals play an important role in referral to physicians for early diagnosis and treatment.

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1 Hailey-Hailey disease (also known as familial benign chronic pemphigus) is a rare, hereditary, autosomal disorder that is distinct from autoimmune-mediated pemphigus. HHD is characterized by blistering in skin folds subject to friction or pressure such as the armpits, groin, neck, under the breasts, and between the buttocks. Unlike true pemphigus, HHD does not affect mucous membranes, and the oral cavity is spared.

2 While not intended as antibiotic prophylaxis for oral procedures, some patients/clients are prescribed antibiotics (with particular anti-inflammatory properties) as part of their pemphigus treatment regimen, including dapsone, tetracycline, and minocycline.

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Oral management implications (cont’d)

- Severe oral erosions interfere with patients’/clients’ proper eating and drinking, as well as hinder nutrient intake. Intravenous (IV) feeding may be needed in very severe cases.
- Oral prophylaxis should ideally be performed prior to the initiation of systemic or topical therapy.
- Routine oral hygiene (including brushing) is often compromised due to pain in the mouth. During the active oral disease stage, frequent patient/client follow-up is recommended (as frequently as every 4 to 6 weeks for debridement), and it should include monitoring for oral candidiasis. During clinical remission, patient/client follow-up care should occur every 6 months.
- The dental hygienist should anticipate that the patient/client may experience pain and bleeding during procedures, and plan for this by scheduling extra time and using suction and gauze as needed.
- Simple hand scaling instruments are often most effective, particularly in patients/clients with severe mucosal disease. Air polishers and harsh abrasives should be avoided due to the fragility of oral tissue. Gentle technique is important.
- Restorative work (such as multiple crowns, implants, and partial or full dentures) is problematic due to tissue damage that can cause extensive ulcerations.
- Disease flares following dental work are fairly common. As result, some authorities recommend prophylactic administration of oral prednisone prior to dental work (e.g., 20 mg/day, in addition to the patient/client’s normal requirement, for 5 to 7 days) for each dental procedure associated with trauma to the gingiva.
- Intraoral photography during appointments can assist in evaluating tissue involvement.
- For multiple oral lesions, corticosteroid mouthwashes are a practical therapy.
- For isolated oral lesions, occlusive topical corticosteroid therapy may be indicated. Custom-made trays can be used to localize steroid application. Topical cyclosporine is also occasionally used.
- For discrete ulcers, intraleSIONal therapy with an injectable corticosteroid (e.g. dexamethasone) may be beneficial. A dentist would typically administer this in conjunction with the patient/client’s physician.
- Numbing lozenges may be used to decrease pain from mouth lesions.
- Patients/clients with oral manifestations should be advised to eat a balanced diet and to avoid rough, acidic, and spicy foods. Alcohol (including in mouth rinses) is contraindicated. During flare-ups, a soft and bland diet is preferred in order to minimize trauma to injured tissue, although this can lead to more plaque accumulation. During periods of remission, patients/clients generally have no dietary restrictions.
- The dental hygienist plays a role in monitoring the patient/client for long-term effects of corticosteroid and immunosuppressive treatment.

Oral manifestations

- Only the vulgaris, vegetans, and paraneoplastic types of pemphigus affect the oral mucosa, with 80% to 90% of patients/clients with pemphigus vulgaris having oral lesions. In most cases of pemphigus vulgaris, oral involvement is severe, and oral lesions are usually the first to appear and last to resolve in any given patient/client.
- All parts of the oral cavity may become involved in pemphigus vulgaris, with the most common areas being the palate, labial mucosa, buccal mucosa, ventral surface of the tongue, and gingiva. Lesions range from shallow ulcerations to fragile vesicles or bullae, which rapidly rupture after formation resulting in painful ulcers. The superficial “ragged” erosions and ulcerations have a haphazard distribution, and they often resemble aphthous ulcers initially.
- Gentle pressure or minor trauma applied to normal mucosa can cause cleavage in the epithelium, which results in blister formation. This is called the Nikolsky sign.

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### Oral manifestations (cont’d)

- Considerable pain occurs with confluence and ulceration of blisters of the soft palate, buccal mucosa, and floor of the mouth.
- Mouth lesions tend to heal more slowly than cutaneous lesions, often taking weeks to several months with the initiation of corticosteroid therapy.
- In paraneoplastic pemphigus (caused by an underlying cancer), very sore and eroded lips are a common presentation.
- Dental caries and periodontal disease occur with increased incidence and severity due to compromised brushing, often coupled with soft diets.
- Candidiasis may occur with the use of oral topical corticosteroids, as well as with systemic therapy.
- Some drugs used to treat pemphigus, such as cyclosporine and gold compounds, have side effects that include bleeding, oral ulcerations, stomatitis, and tender or swollen gums. Gold can also lead to redness and soreness of the tongue. Methotrexate can cause mouth sores, in addition to a sore throat.
- Biologic response modifier drugs, such as tumour necrosis factor inhibitors (e.g., infliximab and etanercept) can cause sore throat and nasal congestion.

### Related signs and symptoms

- Pemphigus vulgaris, accounting for about 80% of pemphigus disease, is a severe, progressive autoimmune disease. Blisters of varying sizes break out on the skin and various mucous membranes, including the lining of the mouth, pharynx, larynx, foreskin of the penis, vagina, anus, and, uncommonly, the conjunctivae. A minority of patients/clients experience only cutaneous lesions, most often on the face, scalp, upper chest, and back, although skin lesions can occur anywhere on the body.
- Pemphigus vulgaris is a relatively rare condition, with annual incidence estimated to range between 1 and 50 per million depending on population characteristics. Males and females are affected equally. While onset may occur in childhood through advanced adulthood, most persons first manifest signs/symptoms between 40 and 60 years of age. Genetic factors are involved, with increased incidence in Ashkenazi Jews and persons of Mediterranean descent. There is no definitive cure.
- Very rarely, certain drugs can cause pemphigus, including penicillamine (a chelating agent) and ACE inhibitors (a class of blood pressure medication).
- All types of pemphigus are characterized by epithelial acantholysis (i.e., breakdown of cellular adhesion between epithelial cells) with intraepithelial vesicle formation.
- Lesions of pemphigus vulgaris typically present as vesicles or bullae, which soon rupture rapidly leaving red, painful ulcers. Skin lesions tend to stay intact longer than mouth lesions, and the blisters are often filled with clear fluid given their intraepithelial nature. Flaccid blisters are more characteristic of pemphigus than of pemphigoid. Ulcers range from small, aphthous-like lesions to large, map-like lesions.
- When skin blisters break, the skin often has a scraped, red, weepy appearance. Widespread rupturing of blisters with consequent ulceration leads to painful debilitation, fluid loss, and electrolyte imbalance. While scarring after blister healing is uncommon, hyperpigmentation of the skin sometimes occurs. Healing of blisters after initiation of corticosteroid therapy usually takes 6 to 8 weeks, but full healing can sometimes take years.

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3 By contrast, pemphigoid lesions result from cleavage of the epithelium from underlying connective tissue at the basement membrane.

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Related signs and symptoms (cont’d)

■ In pemphigus foliaceus, the skin rash tends to have a more scaly appearance (“eczema-like”) rather than blisters, which reflects superficial epidermal involvement. The face is commonly affected, but lesions can also occur on the scalp and other areas of body. Affected skin is itchy or painful, and it is prone to infection.
■ In paraneoplastic pemphigus, pain and redness of the eyes often accompany lip manifestations. The characteristic widespread rash may have areas that blister and become sore, as well as areas that are hive-like and itchy.
■ Sun exposure, trauma, and radiation therapy may exacerbate pemphigus.
■ When untreated, pemphigus vulgaris has a high mortality rate due to dehydration, electrolyte imbalance, malnutrition, and infection. However, the advent of corticosteroid therapy has significantly decreased its life-threatening potential. The mortality rate is now about 8 to 10% within the first 5 years of disease onset (versus nearly 100% before steroids, with 70% fatality within the first year), mostly related to complications of treatment (e.g., systemic corticosteroids) rather than to the disease itself.
■ Osteoporosis, glaucoma, cataracts, type 2 diabetes mellitus, and infections are complications associated with long-term systemic corticosteroid therapy (e.g., prednisone).
■ Other autoimmune diseases may occur in conjunction with pemphigus vulgaris, including lupus erythematosus, rheumatoid arthritis, Hashimoto’s thyroiditis, Sjögren’s syndrome, and myasthenia gravis.

References and sources of more detailed information

■ RDH Magazine, Dentistry IQ Network
■ Canadian Skin Patient Alliance
■ Medscape - Oral Manifestations of Autoimmune Blistering Diseases
  http://emedicine.medscape.com/article/1077969-overview
■ Canadian Pemphigus and Pemphigoid Foundation
■ Canadian Pemphigus
  http://pemphigus.ca/about-pemphigus/
■ International Pemphigus and Pemphigoid Foundation
  http://www.pemphigus.org/research/clinically-speaking/pemphigus/
  http://www.pemphigus.org/research/clinically-speaking/treatments/
  http://www.pemphigus.org/research/clinically-speaking/complications-side-effects/
  http://www.pemphigus.org/research/clinically-speaking/diagnostic-testing/
  http://www.pemphigus.org/research/clinically-speaking/risk-factors/
  http://www.pemphigus.org/research/clinically-speaking/current-consensus-of-disease-outcomes/
  http://www.pemphigus.org/research/clinically-speaking/prevention-and-psychology/

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## References and sources of more detailed information (cont’d)

- Pemphigus & Pemphigoid Awareness Campaign (IPPF)
  
  
  [http://www.pemphigus.org/awareness/for-dental-professionals-students/what-is-pvmmp/](http://www.pemphigus.org/awareness/for-dental-professionals-students/what-is-pvmmp/)
  
  [http://www.pemphigus.org/awareness/for-dental-professionals-students/diagnosis/](http://www.pemphigus.org/awareness/for-dental-professionals-students/diagnosis/)

- Healthline
  
  [http://www.healthline.com/health/pemphigus-vulgaris#Overview1](http://www.healthline.com/health/pemphigus-vulgaris#Overview1)

- Genetic and Rare Diseases Information Center, National Center for Advancing Translational Sciences, National Institutes of Health (U.S.)
  

- Schalock PC, Hsu JTS and Arndt KA (eds.). Lippincott’s Primary Care Dermatology. Philadelphia: Lippincott Williams and Wilkins; 2011. (p. 357 re. prophylactic administration of prednisone prior to dental work in patients with pemphigus)


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

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

** Date: February 16, 2017