Disease/Medical Condition

MUCOUS MEMBRANE PEMPHIGOID
(also known as “MMP,” “cicatricial pemphigoid,” “benign mucous membrane pemphigoid,” and “mucosal pemphigoid”; includes “ocular cicatricial pemphigoid”)

Date of Publication: June 20, 2017

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

- 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 pemphigoid lesions should be referred to a dermatologist, as well as an ophthalmologist for assessment of ocular lesions. Examination by a gastroenterologist is advised for patients/clients with dysphagia or severe oral lesions to detect possible involvement of the esophagus.
  - 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 medical consult advised? .............................................................. See above.

- 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 +/- increased risk of infection (e.g., systemic corticosteroids [i.e., intravenous formulations or oral prednisone], methotrexate, azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil, and biologics [e.g., rituximab 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).

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

- Is medical consult advised? .............................................................. See above.

- 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 +/- increased risk of infection (e.g., systemic corticosteroids [i.e., intravenous formulations or oral prednisone], methotrexate, azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil, and biologics [e.g., rituximab 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

- Gingival tissue is easily injured by food, brushing, and dental hygiene procedures.
- Oral prophylaxis should ideally be performed prior to the initiation of systemic or topical therapy.
- Many patients/clients with MMP have difficulty maintaining good oral hygiene due to pain in the mouth, and therefore they need to be seen often for prophylaxis. During the active oral disease stage, patient/client follow-up is recommended as frequently as every 4 to 6 weeks for debridement, and it should include monitoring for oral candidiasis (particularly if the patient/client is receiving steroid therapy). During clinical remission, patient/client maintenance appointments should occur every 3 to 6 months, with as little disruption to oral tissue as possible.
- Simple hand scaling instruments are often most effective, particularly in patients/clients with severe mucosal disease. Air polishers and abrasive products should be avoided due to the fragility of oral tissue. Gentle technique is important.

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

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Oral manifestations (cont’d)

- 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.
- Because side effects of systemic corticosteroid therapy may outweigh benefits, occlusive high-potency topical steroid therapy\(^2\) is often used instead. A custom-made tray may be used to keep topical medication in place.
- 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.
- Intraoral photography during appointments can assist in evaluating tissue involvement.
- Fastidious oral hygiene, often including chlorhexidine rinses, enhances the effectiveness of topical steroids when gingival involvement is significant.
- The dental hygienist should perform an assessment of at-home oral hygiene practices. Some constituents in dental products may exacerbate oral inflammation — toothpaste and mouth rinses containing sodium laurel sulfate, cinnamon, wintergreen, or spearmint should generally be avoided. A soft bristle brush and antimicrobial rinses can improve tissue response.
- 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

- Oral lesions are the initial manifestation of MMP in 90% of cases.
- The gingiva is the most common site for oral pemphigoid lesions, although bullae, erosions, and ulcers can also occur in other locations in the oral cavity (including buccal mucosa, labial mucosa, palate, and lips).
- Gingival involvement — sometimes described as desquamative gingivitis or gingivosis — often presents as red patches or confluent ulcers extending to unattached gingival mucosa, which cause mild to moderate discomfort. Concomitant erosions may be seen on attached and marginal gingiva. Sloughing and spontaneous gum bleeding are common.
- Bullae are often not seen, because the blisters are short-lived in the mouth.
- The Nikolsky sign\(^3\) can be produced by applying pressure or friction on normal-appearing tissue.
- Over time, the pain associated with chronic oral MMP usually diminishes in intensity.
- Dental plaque accumulation is common due to diminished routine oral hygiene secondary to pain.
- Dental caries and periodontal disease occur with increased incidence and severity due to compromised brushing, often coupled with soft diets.

\(2\) High potency topical steroids include clobetasol, fluocinonide, betamethasone dipropionate, and desoximetasone.

\(3\) Gentle pressure or minor trauma applied to normal mucosa can cause cleavage in the epithelium, which causes bulla formation. This is referred to as the Nikolsky sign.
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Oral manifestations (cont’d)

■ Candidiasis may occur with the use of oral topical corticosteroids, as well as with systemic therapy.
■ Some drugs used to treat MMP, such as cyclosporine, have side effects that include bleeding, oral ulcerations, stomatitis, and tender or swollen gums. Methotrexate can cause mouth sores, in addition to a sore throat.
■ Biologic response modifier drugs, such as tumour necrosis factor inhibitors (e.g., etanercept), can cause sore throat and nasal congestion.

Related signs and symptoms

■ This relatively uncommon disease is predominately seen in adults and the elderly, and rarely in children. Females are affected more commonly than males. There is no definitive cure.
■ MMP is a chronic autoimmune disease involving predominately the oral mucosa and conjunctivae. Other mucosal sites that may be affected include the nasopharynx, larynx, esophagus, and anogenital area. The corneal epithelium may also be affected. Cutaneous lesions are uncommon, and usually occur on the head, neck, and extremities.
■ Lesions result from cleavage of the epithelium from underlying connective tissue at the basement membrane, without the acantholysis found in pemphigus vulgaris. Relative to pemphigus vulgaris, the bullae of MMP are more tense, less fragile, and tend to persist longer.
■ MMP tends to follow a more benign course than pemphigus vulgaris, although lesions often heal with scarring. (This scarring also distinguishes MMP from non-scarring bullous pemphigoid.) Slow spontaneous improvement may be experienced by some patients/clients, whereas others may have a protracted course with episodes of exacerbation alternating with periods of remission.
■ Scar (cicatrix) formation is a particular concern for the eye. Scarring of the canthus and inversion of the eyelashes lead to corneal trauma.
■ Blindness (due to eye involvement), respiratory compromise (due to throat involvement), and infections (due to areas of open skin) are complications. Scar formation in the mucous membranes can lead to significant disability.
■ Voice alterations may occur with laryngeal involvement.
■ Osteoporosis, glaucoma, cataracts, type 2 diabetes mellitus, and infections are complications associated with long-term systemic corticosteroid therapy (e.g., prednisone).

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4 Acantholysis is breakdown of cellular adhesion between epithelial cells, which results in intraepithelial vesicle formation.
5 Many patients/clients with ocular pemphigoid have their eyelashes permanently removed by electrolysis to prevent corneal damage from eyelash rubbing.

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

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3983742/

■ RDH Magazine, Dentistry IQ Network  

■ Canadian Skin Patient Alliance  

■ Medscape - Oral Manifestations of Autoimmune Diseases  
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■ Canadian Pemphigus and Pemphigoid Foundation  

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


* 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