



MUCOUS MEMBRANE PEMPHIGOID

What are the aims of this leaflet?

This leaflet has been written to help you understand more about mucous membrane pemphigoid. It tells you what it is, what causes it, what can be done about it, and where you can find more information.

What is mucous membrane pemphigoid (MMP)?

MMP is the most up-to-date term for this condition. Other names include cicatricial pemphigoid, oral pemphigoid and ocular pemphigoid.

MMP is an uncommon blistering condition which most frequently affects the lining of the mouth and gums. Other moist surfaces of the body (known as *mucous membranes*) can also be affected, and these include the surface layers of the eyes, the inside of the nose, the throat and the genitalia. The skin is sometimes affected by a few scattered blisters. MMP usually starts in middle and old age. Although it is not usually a serious condition in the mouth, the diagnosis of any type of *MMP* is important as it will alert your specialist to the possibility that the condition may involve your eyes, even if you have no symptoms. Eye involvement (known as ocular cicatricial pemphigoid or ocular MMP) does not occur in all people with MMP, but is potentially serious as it may cause scarring and affect your eyesight. Scarring may also affect the throat and the genitalia, and may be very serious if the larynx is affected.

What causes MMP?

The cause of MMP is unknown. It is considered to be an *autoimmune* disease involving antibodies (natural substances important in defending your body against infections) that

react with the surface layer of your mouth (or other mucous membranes) causing blisters, which usually break down to leave ulcers. MMP is not contagious, or due to food allergy.

Is MMP hereditary?

MMP is not an inherited condition.

What are the symptoms of MMP?

- In mild cases MMP may just involve the mouth, causing occasional blisters. These are usually painless until they break to form ulcers that typically cause burning or stinging, especially when eating spicy foods, citrus fruits, drinking hot beverages/alcohol or using mouthwashes that contain alcohol. If the gums are affected, they may feel sore, particularly when brushing teeth.
- In more severe cases, the mouth, throat, eyes, nose, genitalia or skin may also be affected. If your eyes are affected, they may become red and feel gritty or sore.
- Some people with MMP get a stuffy blocked nose and have blood-tinged mucus or nose bleeds, or notice a change in their voice. Occasionally they may experience some difficulty swallowing.
- If the genitals are affected, blisters or ulcers may appear and tend to persist unless treated.
- There may be scattered skin blisters, often on the scalp but occasionally on the face, trunk or limbs.

What does MMP look like?

In the mouth it typically appears as red or ulcerated patches on the inner cheeks, gums or roof of the mouth. Blisters may occur but tend to easily burst and leave ulcers. The gums become red, shiny and ulcerated and may be the only sign of this condition. Active pemphigoid in the eye will usually appear as a red eye and if not treated may lead to scarring. Skin blisters may form crusts and eventually heal with a superficial scar.

How is MMP diagnosed?

MMP cannot be diagnosed solely by its appearance as other conditions can look very similar. One or two small samples of tissue are usually taken by the specialist team (under a local anaesthetic), usually inside the mouth (this procedure is known as a *biopsy*). The diagnosis can then be made by looking at the samples under a microscope and testing them for specific antibodies associated with MMP. A blood test may also be used to detect these antibodies in the circulation.

You may be referred to an eye specialist (ophthalmologist) who can pick up early signs of pemphigoid involving the eyes, an Ear Nose & Throat (ENT) specialist or a dermatologist depending on your symptoms. This multidisciplinary team care is important.

What is the long-term course of MMP?

MMP is usually brought under control with oral medication such as tablets, or injections. Tablets may be needed for several years but in some patients the condition can go into long term remission and eventually medication may be stopped.

How can MMP be treated?

Your doctor will discuss treatment options and help choose the most suitable medication depending on disease severity, your general health and potential medication side effects.

Topical treatments:

Most patients will be prescribed topical treatments for each affected site.

For the mouth:

- *Anaesthetic (analgesic) mouthwashes* are available if your mouth becomes sore and are particularly helpful if used before meals. Benzydamine (Difflam) mouthwash may be helpful.
- *Topical steroids* which can be applied locally to the mouth are helpful for most patients but often require additional oral medication (tablets). These are available as mouthwashes, sprays, pastes and small pellets which dissolve in your mouth. Your specialist may also consider the use of topical steroids inside custom made gum shields. Topical steroids can sometimes cause oral thrush which can be easily treated or prevented with topical antifungals

- If your gums are affected (desquamative gingivitis), it is important that you keep your teeth as clean as possible by regular and effective tooth brushing. If not, a build-up of debris (known as *plaque*) can make your gum condition worse. Your dentist/dental hygienist will be able to give oral hygiene advice and will arrange for scaling of your teeth as necessary.
- An *antiseptic mouthwash or gel* may be recommended to help with your plaque control, particularly at times when your gums are sore. Daily hydrogen peroxide mouthwash (Peroxyl) or occasional chlorhexidine (Corsodyl) twice per week are examples. If possible avoid a mouthwash containing alcohol.

For the skin:

- Corticosteroid creams or ointments are helpful and will be prescribed by your doctor.

For the eyes:

- Treatment will be directed by your ophthalmologist and may include lubricants and topical corticosteroid eye drops although oral medication is needed for most patients. (see below)

Systemic (whole body) treatment:

- In people who are more severely affected with MMP, or when topical treatment has not helped, systemic treatment (taken by mouth or by injection into a vein) may be required usually for several months or years. Your specialist will discuss risks and benefits of different drug options available. Regular blood tests are required, to check for unwanted effects of treatment especially in the first weeks to months of treatment.
- Oral steroids may be prescribed for a few months in high doses, then for longer periods at low doses.
- Other tablets used to treat MMP include dapsone, sulfamethoxypyridazine (SMP), sulfapyridine or other antibiotics such as tetracycline combined with nicotinamide.
- Immunosuppressant drugs such as azathioprine, mycophenolate mofetil, methotrexate, cyclophosphamide or rituximab may be needed for severe MMP. These help MMP by suppressing the body's immune system.

What can I do?

- Avoid spicy, acidic or salty foods if these make your mouth sore.
- Have your mouth checked on a regular basis by a dentist, or oral specialist.
- Keep your teeth clean by using a soft brush and small interdental brushes
- Choose a toothpaste with a mild flavour and free from the foaming agent, sodium lauryl sulphate (SLS).
- Stop smoking and reduce your alcohol intake to recommended limits.
- For the nose, nasal douching can be helpful.
- For the eyes, please seek precise advice from your ophthalmologist.

Where can I get more information about mucous membrane pemphigoid?

Web links to detailed leaflets:

<http://emedicine.medscape.com/article/1062534-overview>

<http://dermnetnz.org/immune/cicatricial-pemphigoid.html>

Links to patient support groups:

Pemfriends: www.pemfriends.co.uk

International Pemphigus and Pemphigoid Foundation: www.pemphigus.org

The Pemphigus Vulgaris Network: <http://www.pemphigus.org.uk/>

For details of source materials used please contact the Clinical Standards Unit (clinicalstandards@bad.org.uk).

While every effort has been made to ensure that the information given in this leaflet is accurate, not every treatment will be suitable or effective for every person. Your own doctor or dentist will be able to advise in greater detail.

This patient information leaflet was written in conjunction with the British Association of Dermatologists (www.bad.org.uk) and the British Society for Oral Medicine (www.bsom.org.uk)

This leaflet has been assessed for readability by the British Association of Dermatologists' Patient Information Lay Review Panel

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