



Patient information: corneal and external disease

Ocular Mucous Membrane Pemphigoid (OMMP)

This leaflet is designed for patients, as well as their relatives, friends and caregivers, to help explain Ocular Mucous Membrane Pemphigoid (or Ocular Cicatricial Pemphigoid as it was previously called).

What is OMMP?

Mucous Membrane Pemphigoid (MMP) is a rare autoimmune disease (a group of diseases caused by the reaction of the person's immune system producing antibodies - known as autoantibodies - which damage normally healthy body substances). These autoantibodies attack the skin and mucous membranes (the soft wet tissue lining the body openings including eyes, mouth, throat, genitals and back passage). When the eyes are affected it is called Ocular MMP (OMMP). For many people with MMP, the eyes are the only site affected by the disease. About 60% of MMP patients have the disease in their eyes as well as other mucous membranes and/or their skin.

In OMMP, the disease creates autoantibodies which damage the conjunctiva. The conjunctiva is the thin mucous membrane which covers the eyeball and lines the inside of the eyelids. These autoantibodies cause conjunctivitis (a red inflamed conjunctiva) and scarring which often results in the eyelashes turning inwards and scratching the eye, making the problem worse. The original name for the disease was cicatricial pemphigoid; cicatricial means scarring. Both eyes are usually affected, although one eye is often worse than the other.

The disease progresses at different rates in different people, with rapid progression over weeks in a minority and slow progression over months or years in most. Early diagnosis in severe cases improves outcomes. With good treatment the effects of the disease can usually be controlled and both comfort and eyesight maintained.

What causes OMMP?

We don't clearly understand what causes OMMP. Although it is known to have a genetic element, it is not an inherited disease and we don't know what triggers the start of the disease. As with other autoimmune diseases, the immune system mistakenly attacks parts of the body. With MMP, your immune system creates autoantibodies which attack the layer immediately under the mucous membranes or skin, causing inflammation and scarring. Blistering is uncommon in the eyes but occurs at other sites. OMMP is not inherited, you can't 'catch' it from someone else and it isn't due to allergies or diet. It usually affects people over 40 (both men and women) but it has been diagnosed in younger adults and, rarely, in children.

How would I know I have OMMP?

In the eyes, the disease starts as conjunctivitis (red, painful and sticky eyes) which may be mild and intermittent or severe and persistent. These symptoms do not improve with treatment by antibiotics or eye drops and can create a lot of pain and irritation. Although scarring in the conjunctiva occurs early, it can be difficult to see with the naked eye until the disease is quite advanced.

As the inflammation and scarring progresses, it can also cause the pocket between the eyelids and eyeball (the conjunctival fornix) to reduce in depth, which may show as a droopy lid or as a smaller gap between the lids.

The eyelids may turn in and the eyelashes may begin to scratch the surface of your eye. This is called trichiasis. The tear producing glands can also be affected by scarring and inflammation, causing the eyes to be dry.

How is OMMP diagnosed and why may this be difficult to diagnose?

MMP is a rare disease and its occurrence in the eyes is, likewise, unusual. This also contributes to the difficulty of obtaining a swift and accurate diagnosis. It is thought to develop in 8 per 10 million people per year. Consequently, this disease is often not the first consideration when a doctor is trying to find a diagnosis.

To confirm a suspected diagnosis of MMP, a small piece of tissue called a biopsy, is taken from the affected mucous membrane and/or the skin under local anaesthetic and tests done to confirm the diagnosis. This test is called a direct immunofluorescence test (DIF). The blood is also tested for the presence of autoantibodies (serology testing).

MMP often starts in the other mucous membranes (most commonly the mouth) or the skin and, if you have an MMP diagnosis, you may be referred to an ophthalmologist by a specialist in oral medicine or a dermatologist. This is to ensure the eyes are monitored for the effects of the disease. If the appearances in the eyes are typical of eye involvement by MMP there is no need for any further tests to confirm the diagnosis.



However, in about a quarter of cases, the eyes are the only or first affected site and the condition of OMMP is diagnosed by an ophthalmologist. The same tests are then done as described above but with the biopsies taken from the conjunctiva of both eyes using anaesthetic eye drops and usually one other site (commonly the mouth or skin).

When these tests are positive, the MMP diagnosis is confirmed. Whereas these tests are usually positive for MMP affecting non-ocular sites (e.g., mouth, nose, skin) they are negative in about 50% of cases when the eyes are the only site affected by MMP; this may result in delays in diagnosis for ocular MMP (OMMP) patients. However, when these tests are negative in OMMP the disease can still be diagnosed by ruling out the other causes of conjunctival scarring and inflammation. In this situation, an opinion by a specialist in OMMP can be very helpful and they will work together with your local ophthalmology service to confirm the diagnosis, provide shared care and optimise your treatment.

Can other parts of my body be affected?

About 60% of MMP patients have some level of eye involvement in their condition. Some people have only the ocular version of MMP, but many people have other mucous membranes such as mouth, nose, larynx, oesophagus and the genital and anus areas affected. It can also cause blistering on the skin. The degree of scarring varies by the location. You are likely to be referred to a variety of medical specialists to monitor the extent of your disease.

How am I going to be treated?

The ophthalmologist will treat cases depending on the severity of disease and the rapidity with which it is progressing. About one in five patients with OMMP have mild disease and don't need specific treatment. However, for most patients specialised treatments are needed to prevent or slow disease progression. Topical treatments (eye drops or eye ointments) haven't been found to be effective in preventing progression of OMMP so the usual treatment is to prescribe drugs taken by mouth (called immunosuppressive drugs). These drugs are designed to reduce the over activity of the immune system that is causing the disease, whilst maintaining it enough to perform its normal activity such as fighting infection. This is quite specialised treatment, very similar to that used for diseases like rheumatoid arthritis.

The immunosuppressive drugs most often used for OMMP are:

- azathioprine
- cyclophosphamide
- dapsone



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- methotrexate
- mycophenolate
- sulfasalazine or sulfamethoxypyridazine
- prednisolone

If your eye disease is particularly severe, a biological treatment such as Rituximab or Truxima might be used, or intravenous immunoglobulin (IViG) which replaces antibodies in your system.

All of these drugs can have serious side effects and you will need to be monitored while you are on them. Since this disease is chronic – which means it might last for a very long time and may require you to take medication indefinitely – you are likely to need regular check-ups.

Where the disease has also caused entropian (the eyelids turning in) and the eyelashes are scratching the cornea, then you may be recommended to have the eyelashes removed with tweezers (epilated) which some patients can do at home, or have surgery either to reposition in-turned eyelids or remove the lashes completely. Treatments to control the other side effects of OMMP may also be prescribed such as lubricating gels, drops or ointments for dry eyes and treatment for blocked eyelid margin glands (blepharitis) that can be treated with hot compresses and lid margin cleaning, as well as keeping your eyelids clean.

Can I be cured?

MMP and, specifically OMMP, are not curable, but with the right treatment at the right level, it is possible to halt the progress of the disease and 'remission' can be achieved. It is vital that there is an early diagnosis and an effective treatment can be found for severe or rapidly developing scarring. At the moment, we are not able to reverse the blindness that very bad or untreated OMMP can cause.

What can I do to manage my disease?

In addition to managing your medications as instructed (and reporting any worrying side effects), you can minimise some of the effects that OMMP has by:

- 1. If you can, remove any eyelashes that are turning inwards or ask someone else to do it. Partners, your hospital clinic, your optometrist and occasionally a GP practice nurse may be able to help.
- 2. Regular treatment of blepharitis for those affected (see the Moorfields blepharitis leaflet)
- 3. Do not let your eyes dry out. Ask for gels or ointments that you can use regularly to keep them moist.



4. This is a chronic severe disease which can be difficult to cope with. The complications need to be understood by your family/friends/employer and you can be helped by others in the patient support groups listed below.

Is there any support I can access?

There are several patient organisations for people with OMMP (and MMP). In the UK there is the PEM Friends support group, found at www.pemfriendsuk.co.uk. There is also a Facebook group, specifically run for patients with MMP. The link to this can be found on the website but we have also put it here for your information:

https://www.facebook.com/groups/121991554546796

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Moorfields Direct telephone helpline

Phone: 020 7566 2345

Monday-Friday, 8.30am-9pm

Saturday, 9am-5pm

Information and advice on eye conditions and treatments from experienced ophthalmic-trained nurses.

Patient advice and liaison service (PALS)

Phone: 020 7566 2324 or 020 7566 2325

Email: moorfields.pals@nhs.net

Moorfields' PALS team provides confidential advice and support to help you with any concerns you may have about the care we provide, guiding you through the different services available at Moorfields. The PALS team can also advise you on how to make a complaint.

