



Pemphigus Vulgaris

What is pemphigus vulgaris?

Pemphigus vulgaris (PV) is a rare autoimmune blistering disease that affects men and women equally, with the average age at diagnosis being 50 years old. Although anyone can develop this condition, some ethnic groups such as Ashkenazi Jews and those of Mediterranean descent are more likely to be affected. The skin and mucous (“wet”) membranes of the mouth, nose, throat, ears and genital areas may be affected. Oral sores may be the first sign of disease with skin blisters developing later.

The most common sites affected in the mouth are the gums, roof of the mouth, inner lips, inside cheeks, and the underside of the tongue. Usually, skin and mouth lesions appear as red and raw areas that are associated with pain and bleeding. Some patients may notice blisters that break down to form sores. Sometimes patients also notice that they have nose bleeds or have developed hoarseness as a result of involvement of those sites.

What causes pemphigus vulgaris?

PV is an autoimmune disease, which means that your own immune system, which normally helps you stay healthy by fighting off infections, becomes confused and attacks your own body by mistake. We do not know why this happens although there may be genes that predispose to this condition. PV is NOT infectious in nature and it cannot be spread to family members or friends.

How do we know it is pemphigus vulgaris?

Diagnosis of PV requires an oral biopsy since other conditions may look similar. A special test called the direct immunofluorescence study will also be performed. This almost always confirms the diagnosis. In some cases, a special blood test called indirect immunofluorescence may also be ordered to help confirm the diagnosis.

How do we treat pemphigus vulgaris?

PV is a systemic autoimmune disease with no known cure. The goal is to control the disease by reducing the formation of blisters and sores and therefore reducing pain. Even if you have only oral disease, you will likely need to be treated with systemic medications to control the disease and keep it from progressing.

You will likely be treated with topical steroids 3-4 times a day for a few weeks. Sometimes if there is a large ulcer, your doctor may recommend treating the area “intralesionally” (with a steroid injection directly into the involved area), to speed the healing process. In severe cases, steroid tablets such as prednisone may need to be taken for several weeks to help heal the lesions.

The most commonly prescribed topical steroids are fluocinonide or clobetasol gel (or compounded clobetasol rinse), and dexamethasone rinse. You may also be prescribed a topical non-steroid medication called tacrolimus, either as an ointment or as a compounded rinse. You may notice what is known as a “black box warning” on the tacrolimus packaging because animal studies showed an increased cancer risk from using this medication. We believe this risk to be minimal compared to the benefits you will experience. In spite of this warning, it is also widely prescribed by dermatologists because it successfully treats many skin conditions.

After symptoms have been brought under control, you may reduce the frequency of therapy to the lowest amount needed to maintain comfort, increasing the frequency during flare-ups as needed. It is a good idea to stop treatment completely if you have no discomfort to let your mouth rest rather than use the topical therapy continuously.



Pemphigus Vulgaris

continued...

Instructions for applying a gel or ointment: After rinsing your mouth with water, gently pat the affected area(s) dry with cotton gauze. Place a small amount of gel on a clean finger, dab it onto the area that hurts and do not eat or drink for 15 minutes for the steroid to be absorbed. It will not hurt you to swallow some of this gel. You may also apply the gel to gauze and place the gauze against the affected area that you are treating. If the gums are involved, a custom tray, like those used for teeth bleaching but covering the affected gums, may be worn with the steroid in it for 30 minutes once or twice a day.

You may notice that the packaging of the steroid may have the following warning: “Not for internal use” or “For external use only.” Such topical steroids have been used for decades to treat inflammatory conditions in the mouth effectively. The warning is there because these steroids are not FDA-approved for this use although there are many studies that demonstrate their effectiveness and safety for treating oral diseases.

Instructions for using a mouth rinse/solution: If you have extensive or difficult to reach oral lesions, you may be prescribed a topical steroid solution (typically dexamethasone) that is used as a mouthwash. A teaspoon (5 ml) of solution should be rinsed for 5 minutes then spat out, and you should not eat or drink for 15 minutes afterwards. It is very important to hold the solution in your mouth for the full five minutes to ensure it works effectively. You may also be prescribed tacrolimus or clobetasol compounded into a rinse by a special compounding pharmacy. You would use it the same way as the dexamethasone.

Any of these topical gels or rinses may cause slight stinging when applied or rinsed. Your doctor may ask you to combine the gel or rinse you are using with topical numbing medicine called viscous lidocaine to ease this burning or stinging sensation. These treatments may cause you to develop a yeast infection (“thrush”) in your mouth. Your doctor may prescribe an anti-yeast (anti-fungal) rinse such as nystatin, clotrimazole troches, or fluconazole tablets to prevent and/or treat the yeast infection. You are particularly susceptible to this if you have diabetes or a history of yeast infections.

Systemic medications

Most patients with PV will ultimately require systemic steroids (prednisone) or other medications such as mycophenolate mofetil, azathioprine, cyclosporine, cyclophosphamide or methotrexate to control disease. Some patients will also require intravenous therapy with rituximab and intravenous immunoglobulin (IVIg). This aspect of your treatment is generally coordinated by a dermatologist, rheumatologist or internist who specializes in this disease. Many patients will continue to require localized topical therapies even if treated with these systemic treatments.

What can I do for comfort?

During flare-ups, avoid crunchy or hard foods (such as toast or chips). Topical numbing agents such as viscous lidocaine may help to reduce sensitivity and discomfort. Viscous lidocaine may be mixed in equal volumes of Benadryl™ and Kaopectate™ or Maalox™ and used as a soothing rinse. Children’s toothpaste (Colgate™ or Crest Kids™), Biotene products or other mild toothpastes with few additives are usually easier to tolerate.

What can I expect?

PV is a chronic condition that may fluctuate in severity. Some days you may have no sores at all, and on other days it may feel worse with more sores. It tends to flare up if your body is stressed, either physically (such as having a cold) or emotionally (such as having work or family problems). Oral sores may lead to difficulties in eating and maintaining good oral hygiene because of pain. One of the most serious complications of this condition is infection of blistered skin; however, the oral lesions are at very low risk of infection. If you feel pain, burning, or other symptoms related to the throat, nose, skin or genital area, you should contact your specialist physician immediately and not wait for your next scheduled appointment.

