CONDITION

Pemphigus vulgaris

Pemphigus vulgaris is an autoimmune condition that causes fragile blisters and painful raw areas on the skin and moist body linings.

WHAT IT IS
Chronic autoimmune blistering condition

BODY SYSTEM
Skin, mouth, and other moist body linings

MAIN SYMPTOMS
Mouth sores, fragile blisters, raw skin, pain

WHAT CAUSES IT
Immune system attacking skin-cell connections

HOW IT IS DIAGNOSED
Skin biopsy, direct immunofluorescence, and blood tests

MAIN TREATMENT
Immune-suppressing medicines and blister care

CAN IT BE CURED?
No - remission is often achievable

IS IT AN EMERGENCY?
Sometimes - with breathing trouble or severe dehydration

LONG-TERM OUTLOOK
Often controllable, but relapses and treatment risks remain

Overview

Pemphigus vulgaris is a long-term autoimmune condition. "Autoimmune" means the immune system mistakenly attacks part of the body. In this case, it attacks proteins that hold skin cells together.

This causes the upper layers of the skin or moist body linings to separate. Fragile blisters form and break easily, leaving painful, raw areas called erosions. The mouth is often affected first. The nose, throat, genitals, anus, eyes, and skin may also be involved.

Pemphigus vulgaris is not contagious. You cannot catch it from another person or pass it through ordinary contact.

Without treatment, widespread blistering can lead to infection, dehydration, and difficulty

eating. Modern immune treatments can usually control the condition, although treatment and follow-up may continue for years.

Symptoms

Painful mouth sores are often the first symptom. They may look like shallow ulcers because the blisters usually break before they can be seen. Sores can occur on the cheeks, gums, tongue, roof of the mouth, or throat.

Mouth involvement may make eating, drinking, brushing your teeth, or wearing dentures uncomfortable. Spicy, acidic, salty, or rough foods may increase the pain without causing the condition itself.

Skin symptoms can include:

The affected skin is often painful rather than itchy. Ordinary rubbing may sometimes cause the surface to peel because the skin-cell connections are weakened.

Sores may also develop inside the nose, throat, genitals, or anus. Eye involvement is less common but can cause redness, pain, light sensitivity, or changes in vision.

Symptoms may begin in one area and later spread. They can improve with treatment and return during a relapse.

- Soft, loose blisters that break easily
- Raw, tender, or crusted areas after blisters burst
- Lesions on the scalp, face, chest, back, armpits, or groin
- Brown or lighter marks after the skin heals

Causes and risk factors

Pemphigus vulgaris develops when the immune system produces antibodies against desmogleins. These are proteins that help neighbouring skin cells stick together. When these connections are disrupted, the cells separate and blisters form.

The exact reason this immune reaction begins is not fully understood. Genetic susceptibility may contribute, but pemphigus vulgaris is not usually passed directly from parent to child. Having a relative with the condition does not mean you will develop it.

It most often begins in middle or later adulthood, although it can occur at other ages. It is reported more often in some ethnic and ancestral groups, but anyone can be affected.

Certain medicines have occasionally been linked to pemphigus or pemphigus-like blistering. This does not mean that everyone taking those medicines is at risk. Do not stop a prescribed medicine on your own. A healthcare professional can review whether a medicine could be contributing and whether an alternative is appropriate.

Pemphigus vulgaris is not caused by poor hygiene, food, stress, or something you touched. Stress may make coping harder and may coincide with symptoms, but it is not the underlying autoimmune cause.

Diagnosis

Several conditions can cause mouth ulcers or skin blisters, so symptoms alone cannot confirm pemphigus vulgaris. Diagnosis is usually made by a dermatologist, a doctor specialising in skin conditions.

A small skin or lining sample, called a biopsy, is central to diagnosis. One sample may be taken from a fresh blister for examination under a microscope. This can show skin cells separating from one another.

Another sample is usually taken from skin next to a blister. It is tested using direct immunofluorescence, a laboratory method that makes immune deposits glow under a special microscope. Pemphigus vulgaris typically produces a net-like pattern around skin cells.

Blood tests can look for antibodies against desmoglein proteins. These tests can support the diagnosis and may help follow immune activity over time, but results are interpreted together with the examination and biopsy findings.

If the mouth, throat, eyes, or genitals are involved, another specialist may help assess those areas.

Treatment and management

Treatment aims to stop new blisters, heal existing erosions, reduce pain, and prevent complications. Because the immune system causes the damage, the main treatments reduce or alter immune activity.

Systemic corticosteroids are medicines that suppress inflammation throughout the body. They can control blistering but may cause important side effects when used for long periods.

Rituximab is an immune treatment that reduces B cells, the cells involved in producing antibodies. It is commonly used for pemphigus vulgaris, sometimes with corticosteroids like

prednisone or hydrocortisone . Other immune-suppressing medicines, such as azathioprine or mycophenolate, may be used to help control the condition or reduce the need for corticosteroids.

Treatment that affects the immune system requires monitoring. This may include blood tests and checks for infection, liver problems, blood-cell changes, bone health, blood pressure, or blood sugar. The checks needed depend on the medicine.

More difficult-to-control cases may require other specialist treatments, such as intravenous immunoglobulin. This contains purified antibodies given through a vein. Treatment choices take account of symptom severity, other health conditions, previous treatment, and pregnancy plans.

Local care remains important even when systemic treatment is needed. It may include:

Do not deliberately break blisters. Open areas should be handled gently because friction can remove more skin. Before vaccinations, dental procedures, or starting new medicines, tell the relevant professional about your condition and immune treatment.

- Corticosteroid gels or rinses for selected mouth lesions
- Non-stick dressings and gentle cleansing for broken skin
- Pain relief that makes eating and mouth care easier
- Dental care and gentle oral hygiene
- Nutrition support when mouth or throat pain limits food intake

Complications

Open skin and mouth erosions can become infected. Warning signs include increasing redness, warmth, swelling, pus, worsening pain, fever, or a bad smell.

Extensive skin loss can lead to fluid, salt, and protein loss. Painful mouth or throat lesions may also cause dehydration, weight loss, or poor nutrition if drinking and eating become difficult.

Skin lesions usually heal without deep scars, but temporary or lasting colour changes can remain. Persistent scalp inflammation or infection may occasionally damage hair follicles.

Immune-suppressing treatment can increase infection risk and cause other side effects. Corticosteroids may affect bones, blood pressure, blood sugar, eyes, mood, muscles, and weight. Other treatments have their own monitoring needs.

Emotional effects also matter. Pain, visible lesions, disrupted sleep, and uncertainty about relapses may affect mood, confidence, relationships, and work.

Prevention

There is no established way to prevent pemphigus vulgaris from first developing. It is an autoimmune condition and is not caused by a preventable infection or everyday behaviour.

Once diagnosed, following the treatment and monitoring plan can reduce the chance that uncontrolled inflammation will cause complications. Avoiding unnecessary rubbing or injury may help protect fragile skin. Gentle dental care can reduce additional irritation and infection in the mouth.

If a particular medicine is suspected of triggering symptoms, any change should be planned with the professional who prescribed it. Stopping some medicines suddenly can be harmful.

Living with it and what to expect

Pemphigus vulgaris usually has a course measured in months and years. Treatment often begins with a period of active control, when the aim is to stop new blisters and allow existing areas to heal. Improvement may take time, particularly when the mouth is involved.

Once symptoms are controlled, medicines may be reduced gradually under supervision. Some people reach remission, meaning there are no new or established lesions for a sustained period. Remission may occur while taking a small amount of treatment or after treatment has stopped.

Relapses can happen. New mouth soreness or a few fragile blisters may be the first signs. Early review can help determine whether the condition is returning or whether there is another cause, such as infection or irritation.

Many people continue their usual relationships, work, exercise, and travel once symptoms are controlled. During active mouth symptoms, softer foods and cool drinks may be easier. During skin flares, loose clothing and non-stick dressings can reduce friction.

Regular follow-up remains important even when the skin is clear. These visits are used to check for subtle symptoms, monitor treatment effects, and decide whether therapy can be adjusted safely.

When to get medical help

Arrange a routine medical assessment if you have persistent, unexplained mouth sores or fragile skin blisters, especially if they are spreading or not healing. These symptoms have many

possible causes and need proper examination rather than self-diagnosis.

If you already have pemphigus vulgaris, contact your care team promptly for new blisters, worsening mouth pain, reduced food intake, or possible treatment side effects. Do not change immune treatment without guidance.

Seek urgent same-day medical help for:

Call emergency services for trouble breathing, inability to swallow saliva, fainting, severe confusion, or signs of a severe allergic reaction. These are emergency symptoms whether or not they are caused by pemphigus vulgaris.

- Fever with spreading redness, warmth, pus, or worsening skin pain
- Difficulty drinking, much less urine, marked dizziness, or other dehydration signs
- Rapidly spreading raw skin or many new blisters
- Eye pain, significant redness, light sensitivity, or vision changes
- Mouth or throat lesions that prevent adequate eating or drinking

Common questions

Is pemphigus vulgaris contagious?

No. It is caused by an immune-system mistake, not by bacteria, viruses, or fungi. Touching a blister does not transmit the condition to another person. An infected open wound may contain germs, but that infection is separate from pemphigus itself.

Can pemphigus vulgaris go away completely?

There is no guaranteed permanent cure, but remission is possible. Some people eventually remain clear without treatment, while others need ongoing medicine or experience relapses. The course cannot be predicted from one early flare.

Is pemphigus vulgaris cancer?

No. It is an autoimmune blistering condition, not cancer. Some treatments also used for cancers, such as rituximab, may be used because they affect specific immune cells.

What can I eat when my mouth is sore?

Soft, bland foods and cool drinks are often easier. Spicy, acidic, salty, very hot, or sharp-edged foods may irritate open areas. There is no specific diet proven to cure pemphigus vulgaris. Ask for nutrition support if pain is causing weight loss or dehydration.

Can I have dental treatment?

Dental care is important, particularly when the gums are affected. Tell the dental team about pemphigus vulgaris and any immune-suppressing treatment. Gentle techniques may reduce injury, and non-urgent procedures may need planning during a severe flare.

Can pemphigus vulgaris be life-threatening?

Severe untreated blistering can be dangerous because of infection, dehydration, and poor nutrition. Immune-suppressing treatments can also cause serious infections. Modern treatment has greatly improved control, but prompt care and long-term monitoring remain important.

Sources

- NHS: Pemphigus vulgaris
- American Academy of Dermatology: Pemphigus-overview, symptoms, causes, diagnosis, and treatment
- British Association of Dermatologists: Pemphigus vulgaris patient information leaflet
- Joly P, Horvath B, Patsatsi A, et al. Updated S2K guidelines on the management of pemphigus vulgaris and foliaceus initiated by the European Academy of Dermatology and Venereology. Journal of the European Academy of Dermatology and Venereology . 2020;34(9):1900-1913
- Murrell DF, Peña S, Joly P, et al. Diagnosis and management of pemphigus: Recommendations of an international panel of experts. Journal of the American Academy of Dermatology . 2020;82(3):575-585.e1
- Didona D, Maglie R, Eming R, Hertl M. Pemphigus: Current and future therapeutic strategies. Frontiers in Immunology . 2019;10:1418