# IPPF Guide to Pemphigus and Pemphigoid

**Information for those living with pemphigus and pemphigoid**

ippf International Pemphigus & Pemphigoid Foundation

# Disclaimer

The information in this guide has been reviewed by the Education and Patient Support working group of the IPPF's Medical Advisory Council. The IPPF does not endorse any drugs, treatments, or products in this guide. Information is provided for informational purposes only. Because the symptoms and severity of pemphigus and pemphigoid vary among individuals, discuss all drugs and treatments with the reader's physician(s) for proper evaluation, treatment, and care.

Information is a critical factor in treating and living with any condition. However, every person's situation is unique. The IPPF reminds everybody to discuss this guide with their doctor or healthcare team to determine if it applies to their situation.

# A Message from our Executive Director

If you are reading this guide, you—or someone close to you—have most likely been diagnosed with pemphigus or pemphigoid. This may be a new diagnosis, or perhaps you've managed your condition for a long time and are now wondering what additional information is available. No matter how you came to the IPPF, I want to welcome you to our community. Though the diseases that bond this community are complex, the IPPF is a place of hope as we work toward fulfilling our mission of improving the quality of life for all people affected by pemphigus and pemphigoid. To this aim, the IPPF exists to bring you accurate, medically-reviewed educational information about pemphigus and pemphigoid diagnosis, management, treatment, and more so that you can live an active and fulfilling life. Though a cure does not yet exist for these diseases, many patients can achieve states of remission with minimal to no disease activity for long periods of time.

At this stage in your disease journey, there's a good chance you are feeling many emotions and worrying about what this all means for your future. This is normal, as it's common for people diagnosed with any chronic illness to experience depression, stress, anxiety, anger, confusion, and/or fear. Trying to understand pemphigus and pemphigoid and their treatments can be overwhelming, not to mention figuring out how factors like diet, sleep, and lifestyle may play a part in your individual experience. With this in mind, I assure you that the IPPF is here to support you on this journey.

This patient guide is intended to provide medically-reviewed information relevant to the most common questions people have when first diagnosed with pemphigus and pemphigoid, as well as educational information about ongoing disease management and treatment options. Through this guide and other IPPF resources, we hope to empower you with the essential knowledge that can make living with pemphigus and pemphigoid much more bearable. Our goal is to help you realize that though your disease may be rare, you are never alone.

In addition to this guide, the IPPF offers several resources on our website: www.pemphigus.org. You can also email us at info@pemphigus.org or call (855) 4PEMPHIGUS.

Sincerely,

Patrick Dunn
IPPF Executive Director

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# Basic Information

Pemphigus and pemphigoid are rare, blistering autoimmune diseases of the skin and mucous membranes. There is currently no cure for either pemphigus or pemphigoid, only remission. Characteristics of pemphigus and pemphigoid include blistering eruptions or lesions on the outer layer of the skin (epidermis) and mucous membranes.

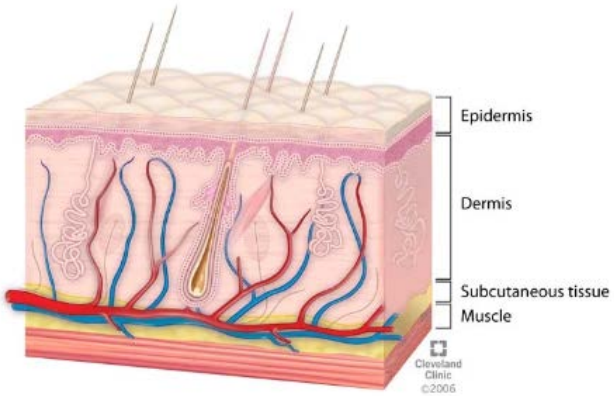
A person's immune system typically makes antibodies to proteins on bacteria and viruses. These antibodies help the rest of the immune system recognize and eliminate the bacteria and viruses and protect the person from infection. However, with pemphigus and pemphigoid, antibodies instead attack healthy proteins in the skin or mucous membranes. These kinds of antibodies are referred to as autoantibodies. Autoantibodies target key intercellular junctions responsible for keeping the skin intact, and when these junctions are disrupted, fluid collects between skin layers, and inflammation can occur, resulting in painful and/or itchy blisters.

## Pemphigus

"Pemphigus" is explicitly used to describe blistering disorders that affect the cells within the upper layers of the skin or mucous membranes. Blisters easily rupture, often resulting in numerous or large patches of denuded skin and mucosa.

## Pemphigoid

“Pemphigoid” is a group of subepidermal blistering autoimmune diseases that affect the skin and mucosa. Here, autoantibodies are directed against the intercellular junctions that connect the epidermis or mucosa to the underlying connective tissue, which can result in tense blisters that fill with fluid. Involvement of the mucosa can sometimes lead to scarring. Sometimes pemphigoid may look like hives or eczema and not have blisters.



If left untreated, pemphigus and pemphigoid may be fatal. Thankfully, corticosteroids such as prednisone and other medications can control pemphigus and pemphigoid, although most deaths now occur from infections, in part due to therapies to treat pemphigus and pemphigoid. The goal of all treatment is to get patients into remission and off of immune suppressing medications. Still, many patients require daily medications or periodic re-treatment to maintain disease control.

## Epidemiology

Approximately 17,000 people in the United States live with pemphigus, and 40,000 live with pemphigoid. Pemphigus and pemphigoid affect men and women approximately equally, although a slight female preponderance has been described for pemphigus and mucous membrane pemphigoid. They are known to affect people across racial and cultural lines. However, there are certain groups of people (Ashkenazi Jews, people of Mediterranean, Asian, Indian, or Persian descent) who have a higher incidence of pemphigus vulgaris. Pemphigus vulgaris is the most common form of pemphigus. However, in some areas of the world, such as Brazil and Tunisia, pemphigus foliaceus is more prevalent.

For pemphigus, disease onset is typically between the ages of 40-60, with the exception of some endemic forms of the disease, which can appear earlier. Paraneoplastic pemphigus due to Castleman's disease also tends to occur in younger individuals. Bullous pemphigoid may appear during pregnancy, or alternatively appears in older aged individuals, with increasing incidence from age 60 onward and even higher incidence in those aged 80 or older.

Blistering in pemphigus and pemphigoid happens because of an immune response resulting in autoantibodies attacking the “glue” that holds mucous membranes or skin cells together. Sometimes, downstream inflammation occurs that can cause itch, pain, and sometimes scarring. Although genetic risk factors have been identified for the development of pemphigus and pemphigoid, these diseases are not directly inherited from parent to child, so it is impossible to predict who may get pemphigus or pemphigoid.

**Pemphigus and pemphigoid are NOT contagious. These autoimmune diseases cannot be passed to others through blood, close contact, or any other means of transmission.**

## Pemphigus Types

The two major types of pemphigus include pemphigus vulgaris and pemphigus foliaceus. IgA pemphigus and paraneoplastic pemphigus are rare forms of pemphigus.

The different forms of pemphigus are distinguished by their clinical features, associated autoantibodies, and laboratory findings.

Hailey-Hailey disease is a genetic disease that can mimic pemphigus, caused by DNA mutation and used to be known as benign familial pemphigus. It is not autoimmune and does not have autoantibodies, so it is NOT considered a subtype of pemphigus.

## Pemphigus Vulgaris (PV)

Pemphigus vulgaris (PV) is the most common type of pemphigus. Autoantibodies most often target desmoglein 3 and/or desmoglein 1, key components of the junctions that hold skin and mucosal cells together, although rare forms of pemphigus that target other autoantigens have been described. The blisters are soft and fragile. In many cases, they may first form in the mouth and then spread to the skin and other mucous membranes such as the nose, throat, genitals, and rarely the lining of the eyelids. Blisters are frequently painful and may sting but are generally not itchy. Blisters in the mouth and throat are painful, making chewing and swallowing difficult and can cause a hoarse voice. PV usually does not cause permanent scarring unless an infection is associated with the sore. PV may also cause nail loss and a change in skin pigment after the blisters heal.

## Pemphigus Foliaceus (PF)

Pemphigus foliaceus (PF) is a more superficial type of pemphigus that only affects the skin, not the mucous membranes. PF is characterized by the loss of intercellular adhesion of skin cells in the upper parts of the epidermis. Blisters may first appear on the scalp and face as itchy and flaky patches, spreading to the chest and back. Autoantibodies attack desmoglein 1. Blisters may be painful and form superficial crusts as they heal. The crusts/blisters can develop over the entire body and can flow together to create large patches of disease activity.

## IgA Pemphigus

IgA pemphigus is characterized by “pimples” or blisters caused by IgA (an antibody) binding to the epidermal cells. IgA pemphigus may resemble pemphigus foliaceus or appear as pustules. The vesicles and pustules are usually, but not always, accompanied by patches of redness and may form a rosette or ring-shaped pattern. The trunk and extremities are common sites for IgA pemphigus. Mucous membranes are usually less commonly affected.

## Paraneoplastic Pemphigus

Paraneoplastic pemphigus, also known as paraneoplastic autoimmune multi-organ syndrome, is often associated with certain forms of cancer. Blisters form inside the mouth and may affect the lungs, leading to a fatal outcome. Sores of the mouth, lips, and esophagus are almost always present, and skin lesions of different types occur. If a cancer diagnosis is not already present, the diagnosis of paraneoplastic pemphigus will prompt doctors to search for a hidden tumor. In some cases, the tumor will be benign. The disease sometimes improves if the tumor is surgically removed.

## Pemphigoid Types

Pemphigoid is a group of subepidermal autoimmune blistering diseases, the most common of which are bullous pemphigoid (BP) and mucous membrane pemphigoid (MMP). In BP, autoantibodies are directed against BP180 and/or BP230, proteins that anchor the basal layer of the epidermis and mucosa to the underlying connective tissue. In MMP, the autoantibody targets may vary across patients. Rarer forms of pemphigoid have also been described in which autoantibodies target other proteins that are involved in anchoring epithelial cells to the connective tissue, such as epidermolysis bullosa acquisita, p200 pemphigoid, laminin 332 pemphigoid, or linear IgA bullous dermatosis.

Pathogenesis and management are quite different for these conditions. Scar formation in mucous membrane pemphigoid can lead to significant disability.

## Bullous Pemphigoid (BP)

Bullous pemphigoid is a subepidermal blistering autoimmune disease that primarily affects the skin. Mucous membrane involvement may occur in 10% to 40% of patients. The disease tends to persist for years, with periods of exacerbation and remission.

The spectrum of presentations is vast, but typically there is an itchy red eruption with tense blisters filled with clear or blood-tinged fluid. Lesions usually appear on the torso and extremities, classically on the legs. Blisters can range in size from a few millimeters to several centimeters and typically heal without true scarring. However, skin discoloration may be left behind after the blisters heal, especially on darker skin. As blisters pop, raw areas of skin known as erosions are revealed, and crusts or scabs can form.

Sometimes blisters do not form, and instead, itchy red bumps and large hive-like areas are the primary skin lesions, sometimes forming an annular pattern. Mucosal (oral, ocular, genital) involvement is also sometimes present, but ocular involvement is rare. BP can be challenging to diagnose in its “non-blistering” stage when just itchy, red areas are visible, as the eruption can easily be confused with a drug reaction.

Localized disease variants have been reported (for example, after surgical procedures), and the prognosis for these variants are often favorable and may be self-limited.

A variant of BP that arises during pregnancy is known as pemphigoid gestationis (PG). In the United States, PG has an estimated incidence of 1 case in 50,000–60,000 pregnancies. PG typically manifests during late pregnancy, typically in the second or third trimester, with an abrupt onset of extremely itchy, hive-like bumps and blisters that often start on the abdomen and can subsequently spread. Disease flares can occur at, or immediately after, delivery. No increase in fetal or maternal mortality has been demonstrated. However, a greater prevalence of premature and small-for-gestational-age babies is associated with PG. PG usually resolves spontaneously within weeks to months after delivery but can reoccur with subsequent pregnancies and rarely can require chronic treatment for disease control.

## Mucous Membrane Pemphigoid

Mucous membrane pemphigoid (MMP) is a chronic autoimmune disorder characterized by blistering lesions that primarily affect the mucous membranes. A subvariant that only affects the eyes and causes scarring is known as ocular cicatricial pemphigoid (OCP).

Oral lesions are often the initial manifestation of the disease, but blisters can affect the nose, throat, esophagus, larynx, genital, rectal, and ocular mucosa. The oral mucous membranes usually heal without scarring, but scarring of other mucosal surfaces can lead to esophageal strictures, urinary or vaginal strictures, conjunctival or laryngeal adhesions that can ultimately lead to difficulty swallowing, urinating, or having sexual relations, and can sometimes lead to blindness and rarely death.

The symptoms of MMP vary among affected individuals depending upon the specific site(s) involved and the progression of the disease. Oral lesions are seen as the initial manifestation of the disease in about two-thirds of the cases. Blistering lesions eventually heal, sometimes with scarring. Progressive scarring may potentially lead to serious complications affecting the eyes and throat.

## Other Forms of Pemphigoid

Linear IgA bullous dermatosis, due to IgA autoantibodies that target protein fragments of BP180, is characterized by rosette-like patterns of blisters and can arise in children and adults. In children, the disease is known as chronic bullous disease of childhood, which is somewhat of a misnomer as the disease typically resolves within months to a few years.

Skin fragility, noninflammatory tense skin blisters, scarring, and healing of blisters with small white cyst-like lesions known as milia characterize epidermolysis bullosa acquisita (EBA). Alternatively, EBA can present as an inflammatory eruption with skin and mucosal lesions reminiscent of bullous pemphigoid. P200 pemphigoid and laminin 332 pemphigoid may be present similarly. The pathogenesis of EBA involves the production of antibodies against type VII collagen, a major component of anchoring fibrils in the basement membrane zones of skin and mucosa, whereas p200 pemphigoid and laminin 332 pemphigoid target other proteins in the basement membrane zone.

The incidence of cancer may be elevated in rare forms of pemphigoid such as EBA and laminin 332 pemphigoid. Age-appropriate cancer screening is recommended.

# Diagnosis

Knowledgeable providers diagnose pemphigus and pemphigoid through special testing and visual exams. At least one immunochemical test to prove the autoimmune nature of the disease is required to confirm the diagnosis. Diagnostic tests include the following:

- **Clinical Presentation:** a visual examination of skin lesions.
- **Lesion Biopsy:** a sample of the blistered skin or mucous membrane is removed and examined under a microscope to determine the layer of skin where blisters occur.
- **Immunochemical tests:**
  - **Direct Immunofluorescence:** a skin or mucous membrane biopsy sample is taken and stained specifically to detect antibodies directly bound to skin tissue. This test is considered the gold standard for the diagnosis of MMP and some of the rarer forms of pemphigoid.
  - **Indirect Immunofluorescence:** a type of blood test that measures circulating autoantibodies in the serum. Indirect immunofluorescence can be performed on salt-split skin to differentiate BP and linear IgA bullous dermatosis (which demonstrate staining on the epidermal side of salt-split skin) from EBA, p200 pemphigoid, and laminin 332 (which demonstrates staining on the base of the salt-split skin). Indirect immunofluorescent testing for MMP is unreliable and is negative in most cases.
- **ELISA**: a serum assay for autoantibodies (anti-desmoglein 1 and desmoglein 3 for pemphigus, and BP180 and BP230 for bullous pemphigoid). A type of VII collagen autoantibody, ELISA is available for the diagnosis of epidermolysis bullosa acquisita but has poor sensitivity to detect disease. Although there is a general correlation between desmoglein 3 and desmoglein 1 ELISA, and disease activity for pemphigus, it is not so in every case, and correlations with disease activity are weaker for BP180 and BP230 autoantibodies and bullous pemphigoid. However, the occurrence of a negative ELISA after treatment generally correlates well with the ability to achieve complete disease remission of systemic immunosuppressive therapies.

# Treatments

The availability of specific treatments may vary from country to country. Medicines may have different names depending on where you live. Please check with your healthcare team or pharmacy to determine which treatments are available where you live or receive treatment.

There are generally two phases to treating pemphigus and pemphigoid:

- **Control/consolidation:** a period of intense therapy, most often with corticosteroids, to suppress disease activity until no new lesions appear. When no new lesions have developed for a minimum of two weeks, and the majority (approximately 80%) of established lesions have begun to heal, most clinicians will taper corticosteroid doses.
- **Maintenance:** chronic therapy with systemic immunosuppressives is often required to maintain disease control. For oral therapies, maintenance dosing represents the lowest dose that prevents new lesions from appearing. For infusion therapies, repeat infusions may be scheduled at regular intervals to prevent disease recurrence.

Prednisone is FDA-approved for pemphigus but not for pemphigoid, even though it is used off-label to treat both disease groups.

Rituximab is FDA-approved for moderate to severe pemphigus vulgaris. Rituximab is not approved for other pemphigus subtypes or pemphigoid, but it is used off-label for many of the pemphigus and pemphigoid disease subtypes. Other anti-inflammatory and immunomodulatory drugs are used off-label to treat pemphigus and pemphigoid, including topical steroids, dapsone, doxycycline, methotrexate, intravenous immunoglobulin, and rarely plasmapheresis or immunoabsorption.

It is essential to recognize that many people require combination therapy of two or more medications. Doctors consider many factors when prescribing medicine, including a person's overall health and well-being, other diseases or conditions, age, cost, and the physician's experience with drugs to treat pemphigus and pemphigoid. Please be sure that you discuss your options with your providers so you can make informed decisions about your healthcare regimen.

It is imperative to make sure that all physicians, doctors, dentists, and specialists involved with a treatment regimen are in contact with one another to avoid conflicting medications and to be sure that each doctor's treatments are working in harmony with the others. All treating doctors should be notified of all prescribed or over-the-counter medications, supplements, and vitamins.

Many treatments available in other countries may have different trade names worldwide. Please check with your healthcare team or pharmacy to determine which treatments are available where you live or receive treatment.

## Corticosteroids

Corticosteroids, even today, are a mainstay of treatment. They mimic the effect of the adrenal hormones naturally produced by your body. Systemic corticosteroids are the most established therapy for managing pemphigus and pemphigoid. In most cases, when used in high doses, they can rapidly control the disease. The most commonly used corticosteroids for pemphigus and pemphigoid include prednisone and prednisolone. Prompt and sufficient doses of corticosteroids are used to bring pemphigus and pemphigoid under control. The amount required will vary depending on the activity and severity of the disease and typically range from 0.5–1.5 mg/kg per day of prednisone for moderate to severe disease. However, doses greater than 0.5 mg/kg per day in older individuals or those with comorbidities that could be adversely affected by high-dose steroids, such as diabetes, should be carefully considered. Once the disease is controlled, steroids may be reduced, which helps mitigate the side effects of these medications. Only stop or decrease the dose of this medication after consulting your prescribing provider.

Topical steroids are also used in the treatment of pemphigus and pemphigoid. Steroid ointments and creams can be applied to the face and body, although chronic use of topical steroids to the skin should be avoided due to the risk of side effects. Steroid ointments, oils, solutions, or foams can be used for the scalp. Many times topical steroids are often more effective for pemphigoid.

Topical treatments of oral lesions include steroid mouthwash, paste, ointment, or gel. Flexible dental trays can help apply topical therapies if the gums are involved.

Corticosteroids have many side effects, some of which can be serious. Please discuss this medication with your healthcare team for a complete list of side effects and considerations to know if this drug is right for you.

## Rituximab (Rituxan®, MabThera, Truxima, Ruxience, Riabni)

Rituximab is an anti-CD20 monoclonal antibody that was FDA-approved in the United States in June 2018 for moderate to severe pemphigus vulgaris in conjunction with short-term prednisone therapy. Rituximab is not approved for any other pemphigus subtypes or any form of pemphigoid in the United States. Rituxan was also approved in Japan in December 2021 for treating refractory pemphigus vulgaris and pemphigus foliaceus. The European Commission approved MabThera in March 2019 to treat moderate to severe pemphigus vulgaris in the European Union. Many clinicians with access to the drug often use it as first-line therapy for moderate to severe pemphigus.

Rituximab targets CD20-expressing B cells for destruction, which reduces the disease-causing antibodies. Rituximab is an immunosuppressant since B cell depletion can impair the ability to fight infections and may prevent full vaccine responses, depending on the length of time between rituximab administration and vaccination.

Rituximab is given as an intravenous infusion through a vein in your arm. Patients receive two “starter” infusions–one on the first day of treatment and another two weeks later. The FDA-approved dose also allows maintenance doses at 6-month intervals for the duration of the treatment regime. Each rituximab maintenance dose is typically 500 mg but can be up to 1,000 mg in case of a disease flare.

Rituximab biosimilars are now available in the United States, including Truxima, Ruxience, and Riabni. Biosimilars have not been expressly FDA-approved for pemphigus or pemphigoid, but are becoming increasingly used by many insurance plans. Please discuss these medications with your healthcare team for a complete list of side effects and considerations.

## Intravenous Immunoglobulin (IVIg)

Intravenous immunoglobulin (IVIg) therapy is prepared by purifying antibodies from human blood donors. IVIg is given intravenously in a vein, typically in your arm. The dosage is weight based, usually administered at a dose of 2 grams per kg of body weight, split across three to five weekdays. It is a collection of healthy antibodies pooled from donors, so it is not immunosuppressive.

Most people tolerate IVIg without problems, although rarely, blood clots and other side effects can occur. Most side effects, such as headaches, occur when IVIg is administered too quickly. Because of this, it is gradually infused, starting at a very slow rate and increasing in intervals until the maximum rate is reached. Please discuss this medication with your healthcare team for a complete list of side effects and considerations.

## Anti-Inflammatory Agents

Anti-inflammatory agents such as dapsone may have a steroid-sparing effect in mild to moderate disease, often in people who are in the maintenance phase but are corticosteroid-dependent. Similarly, doxycycline may have a steroid-sparing effect, particularly for bullous pemphigoid, but it may also be used in pemphigus.

- **Dapsone** is a first-line treatment for linear IgA bullous dermatosis. It can work well for oral manifestations of MMP or mild ocular involvement without evidence of progression in OCP. Patients are often screened for glucose-6-phosphate dehydrogenase deficiency before dosing, typically ranging from 50–200 mg per day.
    - Several risks of dapsone therapy may occur. Please discuss this medication with your healthcare team for a complete list of side effects and considerations.
- **Tetracycline, doxycycline, and minocycline** are commonly used as steroid-sparing agents in the initial and maintenance phase of therapy, often with niacinamide (nicotinamide). Tetracycline is generally administered as 2 g per day with niacinamide, 1.5 g per day (in divided doses). Doxycycline or minocycline are typically dosed at 100 mg twice daily, with or without niacinamide.
    - Tetracycline class antibiotics are generally well tolerated, but rare serious side effects, particularly with minocycline, may occur. Please discuss these medications with your healthcare team for a complete list of side effects and considerations.

## Oral Immunosuppressants

Immunosuppressants are medications used to suppress the immune system and are often used off-label as a steroid-sparing treatment for pemphigus and pemphigoid. The following is a list of commonly prescribed immunosuppressant medications for pemphigus and pemphigoid:

- **Methotrexate:** Dosed weekly (oral or intramuscular injection). Folic acid is typically taken on non-methotrexate days to offset the side effects.

- **Azathioprine (Imuran®, Azasan®):** Typically dosed twice daily. Patients are often screened for thiopurine methyltransferase or nudix hydrolase 15 enzyme activity before dosing, if available.

- **Mycophenolate (CellCept®, Myfortic®):** Typically dosed twice daily. Women of childbearing age must register with the risk evaluation and mitigation strategy (REMS) system due to the potential risk of birth defects in case of unexpected pregnancy.

- **Cyclophosphamide (Cytoxan®):** Due to the potential toxicities, this drug is generally reserved for patients with rapidly progressive OCP as a bridge to rituximab or for those who do not respond to other immunosuppressives.

- **Cyclosporine (Gengraf®, Neoral®, Sandimmune® Capsules, and Sandimmune®):** Cyclosporin is not commonly used for the treatment of pemphigus and pemphigoid in the United States.

All oral immunosuppressants have risks. To determine if these medications are right for you, please discuss this medication with your healthcare team for a complete list of side effects and considerations.

# Clinical Trials

Clinical trials are how many new drugs are developed and approved. Patient involvement is pivotal to the success of these trials.

In a clinical trial, participants receive specific interventions according to the research plan or protocol created by the investigators. These interventions may be medical products, such as drugs or devices; procedures; or changes to participants' behavior, such as diet. Clinical trials may compare a new medical approach to a standard already available one, to a placebo that contains no active ingredients, or to no intervention. Some clinical trials compare interventions that are already available to each other. When a new product or approach is being studied, it is not usually known whether it will be helpful, harmful, or no different than available alternatives (including no intervention). The investigators try to determine the safety and efficacy of the intervention by measuring specific outcomes in the participants. For example, investigators may give a drug or treatment to participants with high blood pressure to see whether their blood pressure decreases.

These studies are critical because they contribute to the overall knowledge and progress of the understanding of pemphigus and pemphigoid. They also play an instrumental role in developing and approving new FDA (Food and Drug Administration) therapies. People who are willing to participate in clinical trials can benefit personally while helping all pemphigus and pemphigoid patients by potentially furthering necessary research.

For more information, check out the IPPF's clinical trial information page:
**www.pemphigus.org/clinical-trial-information/**

# COVID-19

Although the COVID-19 pandemic phase has ended as of 2023, the IPPF is dedicated to our community. The link below is updated as needed with guidance from the IPPF Medical Advisory Council. We recommend contacting your physician with specific questions or concerns about your condition.
**www.pemphigus.org/information-for-pemphigus-and-pemphigoid-patients-related-to-coronavirus-disease-covid-19/**

# Lifestyle

The IPPF recognizes that, in many instances, a patient's life will drastically change from the beginning of symptoms through diagnosis, treatment, and beyond. The following lists are tips collected from various members of the IPPF community.

Information is a critical factor in treating and living with any condition. However, every patient's situation is unique. The IPPF reminds you that any information in this section is meant for educational purposes only and should be discussed with your doctor or healthcare team to determine if it applies to your situation.

## Mental Health

Before the disease is controlled, you might experience difficulty in:

- getting the information you want.
- coping with high drug doses in the initial stages of treatment.
- dealing with frequent outpatient visits.
- coping with erosions and their pain.

**The exact cause of pemphigus and pemphigoid are unknown.**

Some patients find that once pemphigus and pemphigoid are controlled, their lives are not changed too much. Others find that the disease impacts their lives in many ways. In addition to side effects from drug treatment, the impacts can include social or professional relationships and financial implications due to the cost of prescriptions, special dressings, creams, special liquidized food, etc.

Difficulties can be caused by:

- Having to stop work, sometimes permanently, or move into a part-time job.
- Challenges in obtaining disability benefits.
- Lifestyle changes caused by limiting activity to conserving energy.
- Managing the social effects of pemphigus and pemphigoid unpredictability (flare-ups and "bad" days). For example, having to cancel a prearranged social event at the last minute.
- The disfiguring effects of the disease (i.e., weight gain from steroids or visible erosions on the skin that may leave discolored marks that patients often think of as scars).

Understand that you did nothing to cause your illness. How we deal with these life changes makes the difference between a life of coping and moping. Mental health is as important as physical health. Address mental health and any feelings you are experiencing with your healthcare team to find resources.

**Ideas to support patient mental health:**

- Contact your insurance company to find in-network mental health providers.
- Several mental health organizations have trusted databases of licensed providers. Some of the most common are:
  - The American Psychological Association (https://locator.apa.org)
  - The American Association of Marriage and Family Therapists (https://www.aamft.org/Directories/Find_a_Therapist.aspx)
- Local community resources or local support groups to meet your needs.
- Join a support group for persons with chronic illness.
- Online therapy apps:
  - Talkspace (https://www.talkspace.com), Amwell (https://patients.amwell.com), and BetterHelp (https://www.betterhelp.com) are some apps that can offer resources to help you find a therapist.
  - Many people find digital or virtual therapy more convenient, as they do not have to travel.
  - There is also the added benefit of being in your environment and comfort by attending virtual appointments.
  - The Association for Psychoneurocutaneous Medicine North America (APMNA) also has a list of doctors who treat patients with skin diseases https://psychodermatology.us.
- Find hobbies or participate in gentle, physical activities like walking.

If you believe stress is related to increased lesions, addressing and resolving those stress issues is wise. Many causes of stress are not a matter of choice; however, it is possible to change how an individual deals with various sources of stress. One of the most common and effective ways to help reduce stress is to openly and honestly discuss it with a spouse, friend, or therapist. Having someone in your corner helps.


## Nutrition

Many nutrition concerns arise from prednisone or the inability to eat due to mouth lesions. A prompt response with medication is generally prescribed to control a flare of pemphigus or pemphigoid. Here are some ideas to assist with adapting to possible nutrition changes:

- Discuss your current diet, medications, and lifestyle with your physician and/or dietician before changing your diet.
- Consider a referral to a registered dietician who can help you determine the pH of foods, healthy options, and ways to prepare foods that are easy to eat while maintaining nutritional value.
- Consider incorporating protein shakes or fruit/vegetable smoothies into your diet.
- Keep a food journal to help you determine if certain foods are causing a flare.
    - An easy way to do this is to take pictures of your plate before eating and comment in captions on whether blisters appear.

Additional information about nutrition and pemphigus and pemphigoid:

- Prednisone is a drug that requires a diet high in protein and low in carbohydrates, salt, and fat, with particular attention paid to calcium and potassium levels. Calcium with vitamin D supplements is routinely ordered when prednisone is prescribed. If it is not, please inquire if this should be taken.
- Acidophilus, a beneficial bacteria found in yogurt and dietary supplements, is recommended for those who frequently use antibiotics. It also may help prevent yeast infections.
- It has been documented that some patients have sensitivity to garlic, onions, and leeks (the Allium food family). These and other foods may trigger or worsen flare-ups, although many people find stopping these food items does not affect their disease activity.

- Some patients report disease exacerbation by specific food groups containing one or more of four ingredients: thiol, isothiocyanates, phenols, and tannins. However, it is unlikely that the disease will remit based on diet changes alone. Nevertheless, the following ingredients have been implicated by anecdotal reports:
  - Thiols: garlic and other members of the Allium family that contain plants, such as onion, shallot, chive, and leek.
  - Isothiocyanates: abundant in cruciferous vegetables, including mustard, horseradish, winter cress, turnip, broccoli, radish, cabbage, brussels sprouts, and cauliflower.
  - Phenols: Urushiol can cause contact dermatitis and is most notably found in poison ivy, poison oak, and poison sumac, which are related to mango, pistachio, and cashew.
    - The artificial sweetener aspartame is phenolic and common in many food additives.
    - Phenol is in cinnamon, cinnamic acid, and pinene. It is in tomatoes, potatoes, mangos, bananas, milk, and milk products produced when cows consume phenol-laced feed, such as cottonseed.
  - Common sources of tannins are kola nuts, tea, coffee, raspberry, cherry, cranberry, blackberry, avocado, banana, apple, mango, pear, eggplant, grape skins, coffee, cocoa seeds, ginger, ginseng, garlic, rosemary, and arrowroot.

## Itching

- Don’t scratch. Scratching can cause micro-tears in your skin, leading to further lesions or infections.
- Discuss the itch and what it feels like with your doctor openly and honestly.
- Consider using over-the-counter antihistamines or prescription medications.
- Cool compresses can help decrease the itch feeling.
- Wear cotton gloves or mittens while sleeping to keep you from scratching your skin.
- Be sure to drink plenty of water to be appropriately hydrated.
- Moisturize your skin. Ointments are generally more moisturizing than creams; creams are more moisturizing than lotions.

## Skin Care

Excellent skin care is essential, especially when you have a blistering skin disease.

- Ask your doctor for wound care instructions. Taking good care of your wounds can help prevent infection and scarring.
- Ask your healthcare team if you can use topicals and creams on open lesions or skin.
- Minimize sun exposure. Ultraviolet light may trigger new blisters.
- Talk to your healthcare team about the difference between physical and chemical sunscreens and what you should use.
- Minimize trauma to your skin. Avoid situations where your skin could be touched or bumped, such as contact sports, during highly active phases of the disease.
- Discuss good skin care with your healthcare providers. Moisturize your skin using ointments, creams, or petroleum jelly.
- Treat sores and blisters with soothing or drying lotions or wet dressings to ease discomfort, as discussed with your healthcare provider.
- Wear breathable, moisture-wicking fabrics.
- Use a saline spray to keep nasal membranes moist.
- Generously sprinkling talcum powder or Vaseline on your sheets may help keep oozing skin from sticking.

## Bathing

Consider taking baths, not showers. Water from a shower head can spray at a high pressure that can damage, irritate, or remove the skin during the highly active phases of the disease.

- Consider the water temperature. Water that is too hot can be very drying.
- The longer the time spent in the bath, the more drying it can be.
- Use fragrance-free bath products.
- Consider gentle shampoos, such as baby shampoo.
- Liquid shower gels can be more moisturizing than bar soap.
- Blot the skin to dry, or consider air-drying instead of wiping the skin after bathing.
- Applying prescribed ointments or petroleum jelly to damp skin after the shower can help to trap moisture, which is generally beneficial for the skin.

## Dressings

- Consider alternative methods to keep dressings in place, such as using tight t-shirts, camisoles, leggings/tights, Coban dressings, Kling, or tube netting to keep large-area dressings in place instead of tape.
- Discuss tape options with your healthcare providers. Paper tape tends to be less adhesive and damaging to the skin.
- Non-adherent dressings, petroleum jelly dressings, or moistened gauze can reduce the amount the dressing sticks to lesions.
- Discuss the necessity of antibiotic creams/ointments with your healthcare provider.
- Consider a referral to a wound care specialist or homecare nurse to help with dressings and dressing changes.

## Oral Care

- Talk with your dentist about maintaining good oral health. If you have blisters in your mouth, it may be challenging to brush your teeth properly. Ask your dentist what you can do to protect your oral health.
- Brush with a toddler-sized toothbrush. These toothbrushes tend to be very soft, small, and rounded.
- Use children's toothpaste. Consider non-mint/fruit-flavored or flavorless toothpaste with fluoride, as they tend to be less abrasive.
- Increase the number of dental cleanings per year (gentle cleanings every three months) to decrease plaque and build-up in your mouth.
- Use dental tape instead of dental floss. Dental tape tends to be flatter than floss, and dental picks can help keep your teeth free of debris.
- Consider over-the-counter products that keep moisture in your mouth and contain xylitol.
- Focus on brushing one tooth at a time before moving on to a subsequent tooth.

## Eye Care

- Wear sunglasses when outside in the sun. They can help protect you from UV damage and keep dirt and debris away from your eyes.
- Hot compresses can help to open tear-production glands.
- Apply cool compresses to help with pain and itching.
- Use saline eye drops to help keep eyes clear of debris.
- Use preservative-free lubricating drops to keep eyes moistened.
- Use prescription eye drops as directed by your physician.
- Get yearly eye exams to check for disease progression in and around the eyes.
- Have a physician remove eyelashes that turn inward to avoid further trauma and irritation.

# Caregivers, Family Members, and Friends

Although you may be experiencing many changes and stressful situations, remember that caregivers, family members, or friends may also feel some of the same stress. Many caregivers change their usual work schedules to provide as much care as possible; caregiving may be new to them and can bring emotional changes. According to the Caregiver Action Network, 60% of family caregivers are employed and two-thirds have had to adjust their work-life because of their caregiving role.

## Stay Organized

Daily life becomes very hectic when no proper plan is implemented, particularly when the disease is first diagnosed. Keep a plan of action so the journey to remission can run more smoothly.

## Break Time

Caregiving can be stressful, and the constant work may cause fatigue. Let your caregiver take breaks occasionally so that the stress doesn't become a part of their daily routine. There are many resources for caregivers to find the support they need while helping you. A great one is the Caregiver Action Network (http://caregiveraction.org).

## Helping Without Hurting

Often, a caregiver will give up personal time to care for a patient. Speak with your caregiver about including fun activities, like going out to the park and giving your caregiver alone time. You can both feel comfortable and relaxed without having to sacrifice self-indulgence.

# Glossary of Medical Terms

Some terms doctors use may need to be clarified, bringing more questions than answers. Below is a list of words and definitions to help you understand the extensive information a healthcare professional may give you.

## Adrenal cortex:

The outer portion of the adrenal gland, located on top of each kidney. The adrenal cortex produces steroid hormones that regulate carbohydrate and fat metabolism and mineralocorticoid hormones that regulate salt and water balance.

## Antibody:

A blood protein produced in response to and counteracting a specific antigen.

## Antigen:

Any substance that can induce a specific immune response with a specific antibody, specifically sensitized T lymphocytes, or both.

## Autoantibodies:

Antibodies that react with self-antigens (autoantigens) of the organism that produced them.

## Autoimmune disease:

When the body falsely recognizes its tissues as foreign and directs an immune response against them.

## B-cell:

A type of white blood cell derived from bone marrow. B-Cells mature into plasma cells that secrete immunoglobulin (antibodies).

## Cadherins:
A class of type-1 transmembrane proteins involved in intercellular adhesion.

## Corticosteroid:
Any steroid hormone produced by the adrenal cortex that affects carbohydrate, protein, electrolyte metabolism, gonad function, and immune response; any similar synthetic substance used in treating autoimmune, inflammatory, and/or allergic diseases.

## Dermal:
Refers to the middle layer of skin between the epidermis and the subcutaneous fat.

## Dermatitis:
Any inflammation of the skin.

## Dermis:
The second layer of the skin composed of collagen with blood vessels. The dermis is connected to the epidermis by many proteins.

## Desmogleins:
A family of cadherins consisting of DSG1, DSG2, DSG3, and DSG4 proteins. They play a role in forming desmosomes that join cells to one another.

## Epidermis:
The non-vascular outer layer of the skin. It is made up of several layers of cells. The cell-to-cell adhesion in the epidermis is targeted in pemphigus, whereas the adhesion of the epidermis to the underlying gerbil connective tissue is targeted in pemphigoid.

## Glaucoma:
Any of a group of eye diseases characterized by abnormally high intraocular fluid pressure, damaged optic disk, hardening of the eyeball, and partial to complete loss of vision.

## Immunization:
Inducing immunity to an infectious organism or agent in an individual or animal.

## Immunoglobulin:
The technical term for an antibody.

## Incidence:
The probability of developing a particular disease during a given time period; the numerator is the number of new cases during the specified period, and the denominator is the population at risk during the period.

## Intercellular junctions:
Specialized regions on the borders of cells that connect adjacent cells.

## Lesion:
An area of abnormal tissue change.

## Lymphocyte:
The technical term for a type of white blood cell; major subtypes include B lymphocytes and T lymphocytes (also known as B cells and T cells).

## Mucocutaneous:
About or affecting the mucous membranes and the skin.

## Mucous membrane:
Mucous-secreting membranes that line body cavities or open passages to the external environment (also called mucosa). These are the thin, moist coverings of many of the body's internal surfaces.

## Osteoporosis:
A disease in which the bones become extremely porous, are subject to fracture, and heal slowly.

## Pathologic:

1. Indicative of or caused by a morbid condition.
2. About pathology–a branch of medicine that treats the essential nature of the disease, especially the structural and functional changes in tissues and organs of the body caused by the disease.

## Pigment:

A substance that gives color to tissue. Pigments are responsible for the color of skin, eyes, and hair.

## Predisposition:

A susceptibility to a disease/condition.

## Remission:

A decrease in or disappearance of signs and symptoms. In partial remission, some but not all, signs and symptoms have disappeared. In complete remission, all disease characteristics have disappeared, although there is still pemphigus or pemphigoid in the body.

## Risk factor:

A habit, trait, condition, or genetic alteration that increases a person's chance of developing a disease.

## Side effect:

A consequence other than the one(s) for which an agent or measure is used, as the adverse effects produced by a drug.

## Steroid:

Any class of natural or synthetic organic compounds characterized by a molecular structure of 17 carbon atoms arranged in four rings. They include many hormones, alkaloids, and vitamins.

## Steroid therapy:

Treatment with corticosteroid drugs to reduce swelling, pain, and other symptoms of inflammation.

## Subcutaneous:
Beneath the skin.

## Superficial:
Of, affecting, or being on or near the surface.

## Suppression:
The stoppage of a bodily function or a symptom.

## Tissue:
A group or layer of cells that are alike in type and work together to perform a specific function.

## Vulgaris:
Ordinary; of the usual type.

## White blood cell:
A type of immune system cell that helps the body fight infection and disease. White blood cells include lymphocytes, granulocytes, macrophages, and others.

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**For more educational information on pemphigus and pemphigoid, please visit our website at www.pemphigus.org**