

What Is Pemphigus?

Fast Facts: An Easy-to-Read Series of Publications for the Public

Pemphigus is a group of rare autoimmune diseases. Such diseases occur when the body's immune system attacks healthy cells.

Pemphigus causes blisters on the skin and mucous membranes. Mucous membranes are found in the mouth, nose, throat, eyes, and genitals.

Some forms of pemphigus may be fatal without treatment.

What Causes Pemphigus?

Your immune system makes antibodies, which attack viruses and harmful bacteria. With pemphigus, antibodies instead attack healthy cells in the skin or mucous membranes. As a result,

- Skin cells separate from each other
- Fluid collects between skin layers
- Blisters form and may cover a large area of skin.

The cause of this immune system attack is not known. Pemphigus does not spread from person to person. It does not appear to be passed from parent to child. But some people's genes put them more at risk for pemphigus.

Who Gets Pemphigus?

Pemphigus affects people of all races and cultures. Groups at higher risk are:

- People of Mediterranean descent
- Eastern European Jews
- People who live in the rain forests of Brazil.

Men and women both get pemphigus at the same rate. It is most common in middle-aged and older adults. But it can occur in young adults and children.

What Are the Different Types of Pemphigus?

The type of pemphigus depends on where the blisters form.

Pemphigus vulgaris. This is the most common type of pemphigus in the United States. Most cases start with mouth blisters. They can be painful. But most do not itch or leave scars.

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Pemphigus foliaceus. This type most often starts with sores or blisters on the face and scalp. Blisters then show up on the chest and back. It can also cause loose, moist scales on the skin. Most of the sores are itchy, but not painful. This type of pemphigus does not cause mouth blisters.

Pemphigus vegetans. This type causes thick sores in the groin and under the arms.

IgA pemphigus. This is the least harmful type. The blisters look like those in pemphigus foliaceus. This type can also cause small bumps with pus inside. This type of pemphigus is caused by an antibody called IgA.

Paraneoplastic pemphigus. This is a rare type of pemphigus. Special tests may be needed to diagnose it. It occurs in people with some types of cancer and can lead to:

- Painful mouth and lip sores
- Cuts and scars on the lining of the eyes and eyelids
- Skin blisters
- Severe lung problems.

What Is Pemphigoid and How Is It Different From Pemphigus?

Pemphigoid is also an autoimmune skin disease. It leads to deep blisters that do not break easily. Pemphigoid is most common in older adults and may be fatal.

Treatment is much like that for pemphigus. But severe cases may need other treatment.

How Is Pemphigus Diagnosed?

Doctors diagnose pemphigus using:

- A complete history and physical exam.
- A blister biopsy. A sample of a blister is taken off and looked at with a microscope.
- A test where chemicals are put on a skin sample (biopsy). This test is used to find out which type of pemphigus it is.
- A blood sample to measure pemphigus antibody levels.

Pemphigus is a serious disease. The doctor should do all of these tests. No single test is right all the time.

Pemphigus is rare and often the last disease tested for. If you have long-lasting blisters on the skin or mouth, see your doctor. Early diagnosis may allow treatment with only low doses of medicines.

What Type of Doctor Treats Pemphigus?

A dermatologist (an expert in skin diseases) should diagnose and treat pemphigus. For mouth blisters, a dentist can help you maintain good oral health.

How Is Pemphigus Treated?

Pemphigus is treated with medicine. High doses of anti-inflammatory drugs (corticosteroids) are used to control the immune system. They may be taken by mouth or injected. Corticosteroid cream may also be used on blisters.

Other medicines used include:

- Immunosuppressants, which suppress the immune system
- Antibiotics to treat infections.

These medicines can have major side effects. Ask your doctor to tell you about them. Tell each doctor you see about the medicines you take.

If you take corticosteroids, you may need a diet that is:

- Low in fat, calories, and sodium (salt)
- High in potassium and calcium.

It may take months or years for the sores and blisters to go away. Mouth sores heal the slowest, so avoid spicy, hard, and acidic foods.

What Is the Prognosis for People Who Have Pemphigus?

Pemphigus is rarely fatal. Most people with pemphigus can control it with medicines. But pemphigus and its treatments can cause:

- Lost time at work
- Weight loss
- Loss of sleep
- Stress.

The International Pemphigus Foundation provides support services to help people cope with the disease.

What Are Researchers Learning About Pemphigus?

Current research includes study of:

- Risk factors
- Antibodies that attack skin cells
- Better treatments with drugs that have fewer side effects.

For More Information About Pemphigus and Other Related Conditions:

National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) Information Clearinghouse

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The information in this fact sheet was summarized in easy-to-read format from information in a more detailed NIAMS publication. To order the Pemphigus Q&A full-text version, please contact the NIAMS using the contact information above. To view the complete text or to order online, visit www.niams.nih.gov.

For Your Information

This fact sheet may contain information about medications used to treat the health condition discussed here. When this fact sheet was developed, we included the most up-to-date (accurate) information available. Occasionally, new information on medication is released.

For updates and for any questions about any medications you are taking, please contact the U.S. Food and Drug Administration toll free at 888-INFO-FDA (888-463-6332) or visit its Web site at www.fda.gov.