

Pemphigus

Pemphigus is an autoimmune disease, meaning that cells that normally fight infection attack the body itself instead.

There are 2 main forms of pemphigus. One can affect only the skin (*pemphigus foliaceus*). The other can affect either the mucous membranes of the mouth alone or the mouth and the skin (*pemphigus vulgaris*). In most cases, the disease develops on its own, but certain medications also can cause pemphigus to develop.

Pemphigus commonly affects people aged between 40 and 60 years. The disease can last for many years. Treatment may keep the disease inactive or sometimes cure it.

Symptoms

Pemphigus foliaceus causes red spots with crusts and scale on the skin. *Pemphigus vulgaris* causes blisters that break easily, crusts on the skin, and raw, painful sores in the mouth, often on the gums, walls, and roof of the mouth. The skin sites most often affected are the scalp, face, chest, and back. The skin often breaks open as shallow erosions and feels raw and painful. Scars usually do not develop. While the skin can return to almost normal, darker spots may remain after the blisters go away. The mouth often heals later than the skin. You cannot spread pemphigus to other people (it is not contagious).

Diagnosis

Your physician will ask you for information, examine your skin, test the skin strength by rubbing firmly at a spot, take blood, and perform skin biopsies.

Treatment

Treatment involves finding ways to "calm down" the body's immune system. Prednisone, an oral form of steroid, is usually the first treatment used. Some people cannot tolerate the adverse effects of steroids, which can include high blood pressure, weight gain, thinning of the skin, osteoporosis (thin bones), and elevated blood glucose level. Other drugs that take over the effects of steroids on the immune system are sometimes used to allow earlier discontinuation of prednisone treatment. Newer agents are CD20 antibodies such as rituximab, which may provide months of disease relief and reduce the need for prednisone. A drawback of all drugs that calm down the immune system is an increased risk for infections.



FOR MORE INFORMATION

- International Pemphigus and Pemphigoid Foundation
www.pemphigus.org

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