Dermatology Patient Education

Bullous Disease

Bullous (buhl-uhs) diseases are a group of rare blistering diseases. The blisters can form on the skin and mucous membranes. A mucous membrane is the moist tissue lining the mouth, nose, eyes, throat, esophagus (tube that runs from the throat to the stomach), anus, and genitals.

There is no cure for bullous disease. However, treatment can help you manage the challenging symptoms of these types of disease and reduce flares.

WHAT CAUSES A BULLOUS DISEASE?

Bullous diseases occur when the body's immune system mistakes the structures that hold our layers of skin or mucous membranes together as foreign. The body's natural reaction is to attack foreign substances by forming antibodies. When the antibodies attack these substances, the layers of the skin separate and blisters form. You cannot catch bullous disease from someone else.

WHAT DOES A BULLOUS DISEASE LOOK LIKE?

Most bullous diseases include blisters and raw, itchy skin. Before blisters appear, the skin can severely itch and burn. Depending on the type of bullous disease, blisters can last weeks before resolving. The blisters eventually break. The areas then often become raw and sore. New blisters often form where the previous blisters appeared.

When bullous diseases affect the mouth, eating can cause intense pain. Acidic foods, such as oranges and pineapples, can especially make the mouth burn. Even without eating these foods, the mouth sometimes burns. Nosebleeds can suddenly occur when bullous disease affects the lining in the nose.

WHAT ARE THE TYPES OF BULLOUS DISEASE?

Each type of bullous disease has unique traits.

Pemphigus: This disease causes blisters that are soft, limp, and break easily. There are several types of pemphigus, including:

- **Pemphigus vulgaris:** This often begins with painful, raw blisters in the mouth. Blisters also can form anywhere on the body. The blisters break, leaving painful, raw wounds. These are slow to heal. If the disease becomes severe, just rubbing a finger across the skin can blister a large patch of skin. This can be life threatening.

Pemphigus vulgaris

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- **Pemphigus vegetans:** This is a rare form of pemphigus vulgaris. The blisters break, leaving thick, crusty sores. These sores form on the scalp or face, and often where skin touches skin, such as the armpits and groin.
- **Pemphigus foliaceus:** This type of pemphigus begins with sores or blisters on the face, upper trunk and scalp. Blisters can form elsewhere on the body though not inside the mouth. Sometimes this type of pemphigus clears without medical treatment.
- **Paraneoplastic pemphigus:** A very rare form of pemphigus that occurs in people who have a cancerous or benign (not cancerous) tumor. Severe inflammation in the mouth, which can be hard-to-treat, is a common sign of this type of pemphigus.

Pemphigus foliaceus

Pemphigoid: This type of bullous disease causes deep blisters in the skin. These blisters do not break easily. There are different types of pemphigoid, including:

- **Bullous pemphigoid:** This type is common in older adults. Before blisters appear, the person may have itching, a rash, or hives. The itch and blisters can be widespread or in one area on the body. Frequently the itching may be intense. Some people do not get blisters. They develop raw patches of skin or the skin may look like eczema or hives.
- **Mucous membrane pemphigoid:** Also called "cicatricial pemphigoid," this type only affects the mucous membranes. As the blisters and sores heal, scars can form.

Bullous pemphigoid

Linear IgA bullous disease: Blisters that look like a string of pearls often form on the skin. These blisters are extremely itchy. Blisters also may form on the moist tissue that lines the eyes or mouth. This disease occurs both in children and adults, and can be triggered by a medicine.

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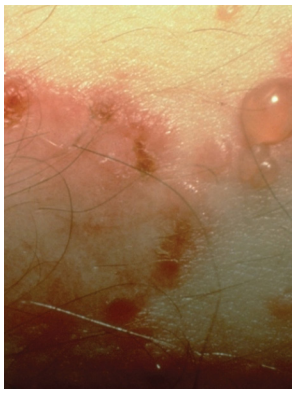

Linear IgA bullous disease

Dermatitis herpetiformis: Blisters, bumps, and patches of raw skin develop that are very itchy. People who have this type of bullous disease are sensitive to gluten, the protein found in wheat, barley, and rye. Some people can prevent outbreaks by not eating food that contains gluten.

Acquired epidermolysis bullosa: Also known as "epidermolysis bullosa acquista," blisters often form on the hands, knees, elbows, feet and ankles. Sometimes blisters cover much of the skin. Blisters also can form on mucous membranes. Many patients experience intense pain. Scars may appear when the blisters heal. The scars can disfigure hands, nails, and other areas of the body.

Epidermolysis bullosa (EB): This group of rare blistering disorders appears at birth or during childhood. These blistering disorders are different than bullous disease because they are not due to the immune system attacking normal structures in the skin. Blisters form in EB because the structures which hold the layers of the skin together are different than skin structures in normal skin. There are many different types of EB depending on how the skin structure is affected. Some types of EB are very severe and lead to extensive scarring, while others are mild.

HOW DOES A DERMATOLOGIST DIAGNOSE BULLOUS DISEASE?

The earlier bullous disease is diagnosed, the more successful treatment can be to control the disease. Without treatment, bullous disease can become disabling and, in some cases, even fatal. A few types of bullous disease increase the risk of certain cancers if left untreated.

To diagnose bullous disease, your dermatologist begins by reviewing your medical history and examining your skin. Different types of bullous disease can look alike. To find out which type of bullous disease you have, lab tests are necessary.

Your dermatologist may remove a blister and some surrounding skin for a biopsy. This is sent to a laboratory for review or your dermatologist will examine it under a microscope. Your dermatologist can perform a biopsy during an office visit which only takes a few minutes to perform. From the same removed skin, special testing is often ordered to determine the type of bullous disease. Sometimes it is also necessary to perform a blood test to diagnose some bullous diseases, in addition to the skin biopsy.

Your dermatologist may take a bit of fluid from a blister. You may not have blisters when you see the dermatologist. If this is the case, a small piece of itchy, raw skin may be removed.

HOW IS A BULLOUS DISEASE TREATED?

Many patients begin treatment by taking a strong oral corticosteroid called prednisone. This medicine quiets the immune system, which may allow the skin to heal and prevent new blisters. Once the disease is under control, prednisone can be gradually stopped. To keep the disease under control after taking prednisone, you may need another medicine. Milder cases can be controlled with topical steroids.

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Some patients cannot take a strong corticosteroid due to side effects. Antibiotics such as tetracycline or erythromycin may also control the disease. Recent studies have shown a combination of antiobiotics and the supplement, niacinamide, have benefited some patients.

Some bullous diseases respond better to powerful steroid-free immunosuppressant medications, such as methotrexate, azathioprine or mycophenolate mofetil. These can be used alone or in combination with each other to treat more severe cases.

Prednisone, dapsone, and the other medications may potentially cause serious side effects. You will need to know the side effects of the medicines you take so that you can alert your dermatologist if problems occur. You also will need to see your dermatologist regularly so that your dermatologist can determine if your treatment is effective, and monitor and adjust your treatment plan.

Bullous disease can be controlled with treatment. If you have bullous disease or have signs and symptoms, you should see a board-certified dermatologist.

Because these diseases can be challenging to live with, you may find that joining a support group for people with bullous disease can help. Your dermatologist may be able to provide you with contact information for a support group in your area.

A board-certified dermatologist is a medical doctor who specializes in diagnosing and treating the medical, surgical, and cosmetic conditions of the skin, hair and nails. To learn more or find a dermatologist in your area, visit aad.org or call toll free (888) 462-DERM (3376).

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