Bullous Pemphigoid

Bullous pemphigoid is a skin disease that causes blisters. It mainly affects people aged over 70. Treatment usually works well to control symptoms. Treatment is usually with steroid creams or medicines, but sometimes other medicines may be used. The condition tends to go away after 1-5 years and then treatment can be stopped. Sometimes the disease persists and treatment is needed long-term.

What is bullous pemphigoid?

Bullous pemphigoid is a skin condition that causes blisters. In some cases the condition becomes severe and serious.

Note: several skin conditions cause blisters and it is important to know exactly which disease you have. They have similar sounding names - for example, pemphigus vulgaris and other types of pemphigus. These different blistering conditions vary greatly in their seriousness, outlook (prognosis) and treatment. For example, see the separate leaflet called Pemphigus Vulgaris (which is generally more serious than bullous pemphigoid).

This photo shows the typical blisters in a 63-year-old man:

![Image of bullous pemphigoid blisters]

Image source: Open-i - see Further reading reference below

Who gets bullous pemphigoid?

Bullous pemphigoid is rare. In the UK it is estimated that between 4-5 people in 100,000 develop it each year. Most people with bullous pemphigoid are aged over 70. It is very rare in children. It can come on in pregnancy, when it is called gestational pemphigoid and requires careful management by a dermatologist and an obstetrician together. Bullous pemphigoid is not infectious and you cannot catch it from an affected person.
What causes bullous pemphigoid?

Bullous pemphigoid is an autoimmune disease. The immune system normally makes antibodies to attack bacteria, viruses, and other germs. In people with autoimmune diseases, the immune system also makes antibodies against a part or parts of the body.

In people with bullous pemphigoid, antibodies are made against the membrane between the top layer of skin (the epidermis) and the next layer (the dermis). This antibody attack causes fluid to build up as blisters between these two layers of skin.

It is not known why bullous pemphigoid or other autoimmune diseases occur. It is thought that something triggers the immune system to attack the body's own tissues. Certain conditions and medications have been linked to bullous pemphigoid, but they are not thought to cause it. They may be trigger factors. They include:

- Diseases such as ulcerative colitis and multiple sclerosis.
- Medications such as furosemide, non-steroidal anti-inflammatory drugs (NSAIDs), captopril, penicillamine and some antibiotic medicines.
- Other factors such as ultraviolet radiation and radiotherapy.

What are the symptoms of bullous pemphigoid?

The first symptoms may be small patches of itchy skin. There may also be a pink rash which can look like eczema. Blisters then develop a week or more later. In some cases, the blisters do not start until months later. The blisters are usually itchy but not painful.

The blisters are quite firm and dome-shaped. The blister fluid is usually clear, but may be cloudy or blood-stained. Any area of skin can be affected, but blisters mostly occur on the arms, legs, armpits and groin. The amount of blistering can vary: sometimes it is just one area, such as the lower leg. In severe cases, the whole body may be affected.

The skin over the blisters is quite strong. It may be several days before the blister bursts. A raw patch of skin is left when a blister bursts, which then heals. Some blisters do not burst; instead the fluid is absorbed into the body and the roof of the blister settles back down on the skin. The blisters usually heal without forming scars.

Some people with bullous pemphigoid get small blisters inside the mouth. The blisters can break to form erosions which look like mouth ulcers.

Bullous pemphigoid is usually itchy. The amount of itch can vary from severe to mild.

How does bullous pemphigoid progress?

If untreated, the blisters and raw areas of skin can cause much discomfort. There is a danger of serious infection occurring on raw areas of skin. Bullous pemphigoid commonly lasts 1-5 years and then often eases or goes away. Future recurrences may occur but these tend to be milder.
How is bullous pemphigoid diagnosed?

The diagnosis may not be clear at first. There are other causes of skin blisters and mouth erosions. Also, the early symptoms (before blisters appear) may look like eczema or allergy. If your doctor suspects that you have bullous pemphigoid, they will usually refer you to a skin specialist. Tests are usually done to confirm the diagnosis. These are:

- **A small sample (biopsy)** of skin may be taken. This is looked at under the microscope and tested to confirm that the blisters are due to bullous pemphigoid.
- **A blood test** can detect the antibody that causes bullous pemphigoid (the bullous pemphigoid auto-antibody). The antibody may also be detected in urine or in blister fluid.

What is the treatment for bullous pemphigoid?

Treatment involves suppressing the immune process, so that itching and blistering are reduced. The aim is to find the right balance of treatment. You need to feel comfortable and not have many blisters, yet without too many side-effects of treatment. Usually the best balance is the point where there may be a few blisters or symptoms which you can tolerate. This may be better than totally suppressing all symptoms, which could mean using large amounts of treatment and having more side-effects. Treatments used for bullous pemphigoid are:

**Steroids**

Steroid creams, also called topical steroids, are a safe and effective treatment for bullous pemphigoid. High-strength steroid creams (for example, clobetasol) will normally be needed. Research suggests that steroid creams are a good option for most types of bullous pemphigoid, even the more severe forms. This research also suggests that certain steroid creams are more effective than steroid tablets, have fewer side-effects and give better results overall.

Steroid tablets such as prednisolone are also used as treatment for bullous pemphigoid. They are used when the rash is widespread, or there are practical problems applying the cream. Steroids reduce inflammation and suppress the immune system. The dose of steroid depends on how severe the bullous pemphigoid is. A medium or high dose is usually needed at first, and the dose can then be reduced once the blisters clear up. The aim is to find the lowest dose needed to control symptoms, which varies from person to person.

Steroid creams and tablets can be used together, and this may mean that a lower dose of tablets can be used. Treatment is usually needed for between six months and four years. Bullous pemphigoid tends not to return after that.

All steroids, whether tablets or creams, can have side-effects. Side-effects from steroids can sometimes be serious, especially if you take high-dose steroids for a long time. For example, you are more prone to certain infections if you take regular high-dose steroids. Another side-effect is ‘thinning’ of the bones (osteoporosis). If you take steroid treatment for more than a month, you will probably be advised to take vitamin D and calcium tablets to help prevent osteoporosis. You should be aware that long-term steroid treatment should not be stopped suddenly. See the separate leaflet called Oral Steroids for more details.

**Other treatments**

There are some other treatments which can help with bullous pemphigoid. These are:

- Skin treatments such as dressings and wound care, which will be needed if you have areas of raw skin.
- Medicines called dapsone and sulfonamides, which are sometimes used if steroids have not been effective. They tend to cause more side-effects and so are not used as often as steroids.
- A newer medication called rituximab, which has been useful in difficult-to-treat cases.

What is the outlook for bullous pemphigoid?

The outlook (prognosis) is generally good. Bullous pemphigoid often goes away after 1-5 years. Meanwhile, treatment usually keeps the blisters away or down to a tolerable level. Often, treatment can be stopped after about 1-5 years, as it is no longer needed.

Bullous pemphigoid can sometimes cause serious illness, which may cause death. This is because:

- Serious infection of raw skin is dangerous. Bullous pemphigoid most commonly affects older people, who are more prone to develop serious illness if a skin infection occurs.
- Side-effects from steroids may be a problem and can sometimes be serious.

Further reading & references

- Guidelines for the management of bullous pemphigoid; British Association of Dermatologists (2012)

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