

## **What can we tell you about PEM?**

(From a non-expert perspective)

Pemphigus and Pemphigoid (as well as some related diseases) are groups of auto-immune diseases that affect the skin and/or mucous membranes. They are also grouped under the title auto-immune bullous diseases (AIBD).

Like other auto-immune diseases, they occur when your immune system starts to attack parts of the body that it shouldn't and, in our case, that is your skin. Sometimes the blisters start in your mouth. The antibodies which are a consequence of your faulty immune activity attack the glue which holds the layers of your skin (the dermis and epidermis) together and this, in turn, creates blisters. Although quite often, the early stages of Bullous Pemphigoid appear like a rash and do not always erupt into blisters.

Differences in the various blistering diseases are created by the way the immune system attacks the various levels of the dermis and epidermis.

No-one knows what causes PEM. We do know that, like many other related diseases, there is a genetic fault at the start, and that usually a trigger such as stress, another illness or a drug used to treat another condition that causes the PEM to erupt. It is possible also, that the fact that the immune system weakens with age, that these diseases tend to appear in later years.

The PEM diseases are generally very rare, although the symptoms can easily be confused with other skin ailments. That causes problems with finding a diagnosis and treatments are usually from the range of borrowed drugs – drugs that have been “repurposed” from other similar inflammatory or auto-immune diseases.

Pemphigus and Pemphigoid can be disfiguring but are not infectious and cannot be caught or passed on to anyone else.

A cure for these diseases is unavailable today, but they can be treated successfully, and you can achieve remission from them, with either no ongoing treatment or a very small maintenance dose of the drug that manages your PEM.

To have a confirmation of the Pemphigus or Pemphigoid, there are several tests.

In Pemphigus, blisters tend to form when the skin is rubbed (Nikolsky sign).

An ELISA test is an enzyme linked immunosorbent assay also called that detects and measures antibodies in your blood. It involves taking a blood sample.

The most reliable test is an indirect immunofluorescence test which shows if and where the layers of the dermis or epidermis are being targeted. This test involves taking a biopsy from a blister.