IMMUNE Pharmaceuticals | ippf International Pemphigus & Pemphigoid Foundation

Bullous Pemphigoid (BP)

Bullous Pemphigoid (BP) is an acquired autoimmune skin blistering disease. It is a very rare disease that usually occurs in the elderly, but is actually the most common of the autoimmune skin blistering diseases. BP affects a lower layer of the skin, between the epidermis and the dermis, creating blisters that do not break easily. Bullous Pemphigoid typically involves the skin, with blisters presenting predominantly on the abdomen, groin, back, arms and legs, but occasional patients may have involvement of the mucous membranes. The blisters may itch and be painful.

For a definite diagnosis, doctors should consider:

Clinical Presentation: Visual examination of skin and mucous membrane lesions.

Lesional Biopsy: A sample of the blistered skin is removed and examined under the microscope.

Direct Immunofluorescence Biopsy: A sample of normal, uninvolved skin is removed and examined for the presence of antibodies in the skin.

Indirect Immunofluorescence (IIF) / ELISA: These tests measure the presence of autoantibodies circulating in the blood. IIF is non-specific and measures the presence of antibodies in the blood, while the ELISA assay is specific and can detect the presence of BP 230 and/or BP180 antibodies.

Patient Support

The International Pemphigus & Pemphigoid Foundation (IPPF) is a patient organization devoted to providing support, relief and help to thousands of pemphigus and pemphigoid patients and their families around the world through peer support, public and professional awareness, and patient education programs. The 2016 global theme, "Patient Voice", recognizes the crucial role patients play in voicing their needs and in initiating change that improves their lives and the lives of their families and caregivers. This year's Rare Disease Day events are planned in all 50 states, and in more than 80 countries. Immune Pharmaceuticals and the IPPF announce their support of Rare Disease Day and their joint continued effort to raise awareness of bullous diseases, including Bullous Pemphigoid.

Other Facts

- Bullous Pemphigoid normally, but not invariably, responds to systemic corticosteroids (alone or combined with other oral agents), with most patients improving on prednisone at a dosage of 80 mg/day or less but there is no consensus on optimal dosing.
- High-potency topical corticosteroids are also commonly used for the management of BP, and may be used as monotherapy for patients with mild disease.
- ~70% of patients treated with systemic corticosteroids suffer from toxicities and major short and long-term side effects including but not limited to diabetes, glaucoma, peptic ulcers, skin atrophy and psychosis.
- Because of this toxicity profile of systemic corticosteroids, there is tremendous need for new and better therapeutic options.
- Eotaxin-1 is a pro-inflammatory protein, which is believed to play a significant role in Bullous Pemphigoid. Eotaxin-1 levels are correlated with disease severity and are elevated in blood and blister fluids of moderate-to-severe BP patients.
- Bertilimumab is an experimental drug targeting eotaxin-1 that has been approved by the FDA for clinical trials — to learn more about clinical trial centers please contact Immune Pharmaceuticals (www.immunepharma.com) or the IPPF (www.pemphigus.org).