

# Bullous Skin Diseases



Disease

**Bullous Pemphigoid**

**Pemphigus Vulgaris**

**Epidermolysis Bullosa**

Presentation

- The classical lesion is a 1 to 3 cm, tense bullae on an erythematous, urticarial, or noninflammatory base.
- Blisters may be numerous and widespread.

- Painful blisters and erosions on the skin and mucous membranes.
- Nikolsky sign: Induction of blistering via mechanical pressure at the edge of a blister or on normal skin.

- Commonly presents as trauma- or friction-induced blistering mainly limited to the palms and soles.
- Lesions often heal with postinflammatory hyperpigmentation.

Location

The trunk, extremity flexures, axillary folds and inguinal folds are common sites for cutaneous involvement. Mucosal lesions are present in 10 to 30% of patients.

First presentation is normally on the mucous membranes, followed by cutaneous manifestation on the upper chest, back, scalp, and face.

These may occur anywhere on the body, but most commonly appear at sites of friction and minor trauma such as the feet and hands.

Comorbidities

Often seen in patients with neurological diseases, particularly stroke, dementia, and Parkinson's disease.

Often accompanied by secondary skin infections and nutritional deficiencies due to difficulty eating.

Often accompanied by nutritional compromise mediated by feeding difficulties due to recurrent mucosal lesions of the oropharynx, esophagus, and small intestine.







Population

Often presents in people over 80 years of age and mostly affects people over 50. It can occur in younger adults, but Bullous pemphigoid in infants and children is rare.

Higher prevalence among individuals with Jewish ancestry and inhabitants of India, Southeast Europe, and the Middle East. Pemphigus usually occurs in adults, with an average age of onset of 40 to 60 years.

Inherited disease. Usually occurs at birth or shortly after.

# Bullous Skin Diseases Continued

Disease	Bullous Pemphigoid	Pemphigus Vulgaris	Epidermolysis Bullosa
Causes	<p>Autoantibody deposition at the epithelial basement membrane zone. Association with human leukocyte antigen (HLA). Can be triggered by medications and infection.</p>	<p>Characterized by acantholysis (loss of keratinocyte to keratinocyte adhesion), which is induced by the binding of circulating immunoglobulin G (IgG) autoantibodies to intercellular adhesion molecules.</p>	<p>Heterogeneous inherited skin fragility disorder characterized by disruption at the dermoepidermal junction or in the basal layer of the epidermis</p>
Treatments	<ul style="list-style-type: none"> <li>• <b>High potency topical corticosteroid:</b> First-line option in most cases. Lower mortality and complication rate compared to oral prednisone.</li> <li>• <b>Oral corticosteroid:</b> Useful for rapid onset of resolution.</li> <li>• <b>Doxycycline:</b> Less often used, but also an effective option.</li> </ul>	<ul style="list-style-type: none"> <li>• <b>Systemic corticosteroid:</b> Prednisone is the mainstay of medical treatment for controlling the disease. IV methylprednisolone may be required to achieve control in some cases.</li> <li>• <b>Rituximab:</b> Can be used alongside prednisone in severe cases.</li> <li>• <b>Other immunosuppressants (MMF, AZA, MTX):</b> for refractory cases.</li> </ul>	<ul style="list-style-type: none"> <li>• Symptomatic treatment to protect the skin and stop blister formation.</li> <li>• Avoiding activities that cause skin friction.</li> <li>• When GI tract is involved, soft diet and stool softeners.</li> <li>• Pierce, drain and dress blisters to promote healing by wound care professionals.</li> </ul>
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## References

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## Image Citations

1. Oakley A. Bullous pemphigoid. Accessed November 20, 2023. <https://dermnetnz.org/topics/bullous-pemphigoid>
2. Ngan V, Oakley A, Shakeel M. Pemphigus vulgaris. Accessed November 20, 2023. <https://dermnetnz.org/topics/pemphigus-vulgaris>
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