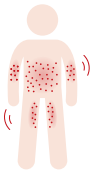
# Bullous Pemphigoid (BP): Don't Miss the Diagnosis



## Clinical Presentations[1-10]

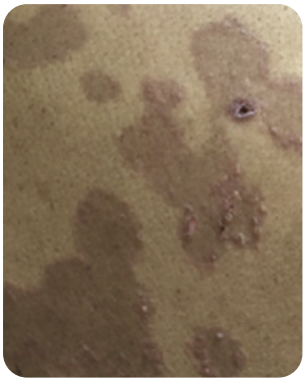
**BP is a chronic relapsing autoimmune skin disease characterized by intense itch, inflammatory lesions, and blister formation**

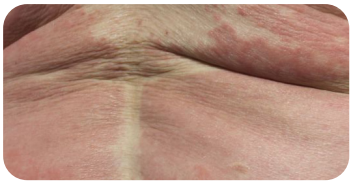
### Non-bullous presentation/phase*

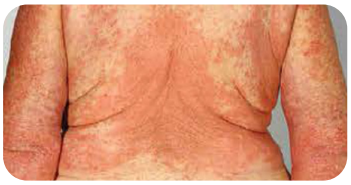
Intense itch with or without inflammatory skin lesions (ranging morphologically from urticarial to eczematous). Occurs in up to 20% of cases.

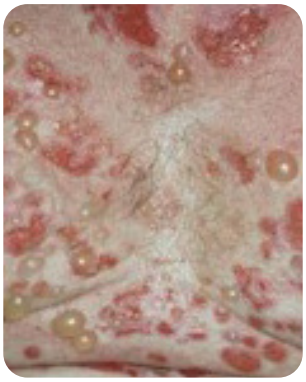
### Bullous presentation/phase

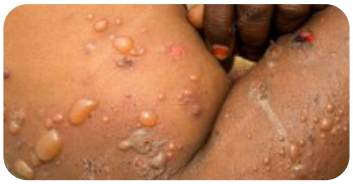
Intensely pruritic bullous dermatosis, with blisters usually arising from apparently normal or erythematous skin. Mucosal involvement reported in 10-30% of patients

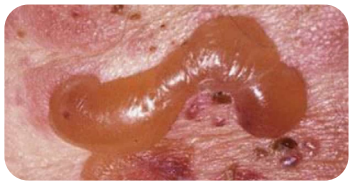












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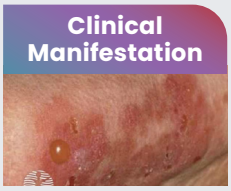
## Diagnosis[5,9-15]

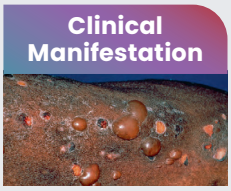
### Clinical assessment

- Medical history
- Patient-Reported symptoms
- Physical examination

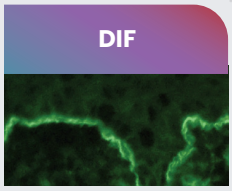
### Laboratory work

- Skin biopsy sample
  - Histopathology
  - Direct immunofluorescence (DIF)
- Serum sample
  - Enzyme-linked immunosorbent assay (ELISA)
  - Indirect immunofluorescence (IIF)

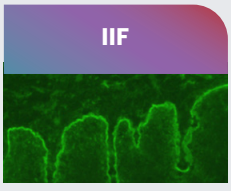








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**BP has polymorphous presentations with a wide spectrum of non-bulbous and bullous manifestations. As such, diagnosis can be challenging, and a broad differential diagnosis should be considered.[10,16,17]**

*May persist for several weeks or months before the formation of bullae, or it may remain non-bullous indefinitely in atypical variants.

BP, bullous pemphigoid.

**References: 1.** Schmidt E, Zillikens D. *Lancet.* 2013;381:320-332. **2.** Zeidler C, et al. *Front Immunol.* 2019;10:1303. **3.** Di Zenzo G, et al. *Clin Dermatol.* 2012;30:3-16. **4.** Kridin K, Ludwig RJ. *Front Med (Lausanne).* 2018;5:220. **5.** Tamzazian S, Simpson CL. *JAAD Case Rep.* 2020;6(11):1173-1178. **6.** PCDS. Accessed September 13, 2024. https://www.pcds.org.uk/clinical-guidance/bullous-pemphigoid1 **7.** Science Photo Library. Accessed September 13, 2024. https://www.sciencephoto.com/media/1227663/view **8.** Schmidt E, et al. *Dermatol Clin.* 2011;29:427-438. **9.** DermNet New Zealand. Accessed September 13, 2024. https://dermnetnz.org/topics/bullous-pemphigoid-images **10.** Schmidt E, Groves R. Immunobullous diseases. In: Griffith C, Barker J, Bleiker T, Hussain W, Simpson RS (Eds). *Rook's Textbook of Dermatology.* Chichester. Wiley-Blackwell; 2024:50.1-50.61. **11.** Borradori L, et al. *J Eur Acad Dermatol Venereol.* 2022;36(10):1689-1704. **12.** van Beek N, et al. *Expert Rev Clin Immunol.* 2016;12:267-277. **13.** Witte M, et al. *Front Med.* 2018;5:296. **14.** James W, Elston D, Treat J, et al. Andrews' Diseases of the Skin: Clinical Dermatology. 13th ed, 2019; Elsevier. **15.** Hammers CM, Stanley JR. *Annu Rev Pathol.* 2016;11:175-197. **16.** Pratasava V, et al. *Medicina (Kaunas).* 2021; 4;57(10). **17.** Miyamoto D, et al. *An Bras Dermatol.* 2019;94(2):133-146. **18.** Lu L, et al. *J Cosmet Dermatol.* 2022;21(10):4818-4835. **19.** Persson MSM, et al. *Br J Dermatol.* 2021;184(1):68-77. **20.** Kouris A, et al. *An Bras Dermatol.* 2016;91(5):601-603. **21.** deGrazia TM, et al. *J Invest Dermatol.* 2019;139:b14. **22.** Kluger N, et al. *Clin Cosmet Invest Dermatol.* 2020;13:73-76. **23.** Han A, Zeichner JA. *J Clin Aesthet Dermatol.* 2009;2(5):19-28. **24.** Penha MA, et al. *An Bras Dermatol.* 2015;90(2):190-194. **25.** Venning VA, et al. *Br J Dermatol.* 2012;167(6):1200-14. **26.** Le ST, et al. *JAMA Dermatol.* 2024;160(1):107-109. **27.** Langan SM, et al. *J Invest Derm.* 2011;131:631-636. **28.** Huttelmaier J, et al. *Front Immunol.* 2023;14:1196999. **29.** Kibsgaard L, et al. *Acta Derm Venereol.* 2015;95:307-311. **30.** Lee J, et al. *Front Immunol.* 2019;10.

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# Disease Burden in Bullous Pemphigoid

## Epidemiology[6,10,18,19]

BP mostly affects the elderly

Incidence is increasing and is estimated to be up to 76.3 cases/million*

Prevalence is increasing, with estimated range of 0.13-1.13%

## Burden of Disease[11,20-25]

### Physical burden

- Physical discomfort (lesions, itch, pain)
- Sleep disturbance
- Fatigue
- Physical limitations
- Dyspigmentation
- Increased risk of infection

### Treatment burden

- Toxicities/adverse events and increase mortality risk due to corticosteroids and other systemic treatments
- Frequent lab monitoring
- Drug interactions
- Frequent topical/oral drug administration


### Mental health burden

- Depression
- Loneliness
- Embarrassment
- Anxiety

### Socioeconomic burden

- Greater dependence on caregivers
- Increased healthcare resource utilization due to repeated follow-up visits, lab monitoring, treatments, and hospitalizations

**There remain unmet needs in the management of bullous pemphigoid[26]**

## Additional Burden of Comorbidities[1,10,27-29]

**BP is typically associated with serious chronic comorbidities and can be life threatening**

- Neurologic Disease[10,27,28]
- Metabolic Disease[10,28,29]
- Cardiovascular Disease[10,28,29]
- Liver and Kidney Disease[28]
- Malignancy[10,28,29]

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*Incidence is estimated to be 10 cases/million in the US.[30]
BP, bullous pemphigoid; QoL, quality of life.

**References: 1.** Schmidt E, Zillikens D. *Lancet*. 2013;381:320-332. **2.** Zeidler C, et al. *Front Immunol*. 2019;10:1303. **3.** Di Zenzo G, et al. *Clin Dermatol*. 2012;30:3-16. **4.** Kridin K, Ludwig RJ. *Front Med (Lausanne)*. 2018;5:220. **5.** Tamzazian S, Simpson CL. *JAAD Case Rep*. 2020;6(11):1173-1178. **6.** PCDS. Accessed September 13, 2024. https://www.pcds.org.uk/clinical-guidance/bullous-pemphigoid1 **7.** Science Photo Library. Accessed September 13, 2024. https://www.sciencephoto.com/media/1227663/view **8.** Schmidt E, et al. *Dermatol Clin*. 2011;29:427-438. **9.** DermNet New Zealand. Accessed September 13, 2024. https://dermnetnz.org/topics/bullous-pemphigoid-images **10.** Schmidt E, Groves R. Immunobullous diseases. In: Griffith C, Barker J, Bleiker T, Hussain W, Simpson RS (Eds). *Rook's Textbook of Dermatology*. Chichester. Wiley-Blackwell; 2024:50.1-50.61. **11.** Borradori L, et al. *J Eur Acad Dermatol Venereol*. 2022;36(10):1689-1704. **12.** van Beek N, et al. *Expert Rev Clin Immunol*. 2016;12:267-277. **13.** Witte M, et al. *Front Med*. 2018;5:296. **14.** James W, Elston D, Treat J, et al. Andrews' Diseases of the Skin: Clinical Dermatology. 13th ed, 2019; Elsevier. **15.** Hammers CM, Stanley JR. *Annu Rev Pathol*. 2016;11:175-197. **16.** Pratasava V, et al. *Medicina (Kaunas)*. 2021; 4;57(10). **17.** Miyamoto D, et al. *An Bras Dermatol*. 2019;94(2):133-146. **18.** Lu L, et al. *J Cosmet Dermatol*. 2022;21(10):4818-4835. **19.** Persson MSM, et al. *Br J Dermatol*. 2021;184(1):68-77. **20.** Kouris A, et al. *An Bras Dermatol*. 2016;91(5):601-603. **21.** deGrazia TM, et al. *J Invest Dermatol*. 2019;139:b14. **22.** Kluger N, et al. *Clin Cosmet Invest Dermatol*. 2020;13:73-76. **23.** Han A, Zeichner JA. *J Clin Aesthet Dermatol*. 2009;2(5):19-28. **24.** Penha MA, et al. *An Bras Dermatol*. 2015;90(2):190-194. **25.** Venning VA, et al. *Br J Dermatol*. 2012;167(6):1200-14. **26.** Le ST, et al. *JAMA Dermatol*. 2024;160(1):107-109. **27.** Langan SM, et al. *J Invest Derm*. 2011;131:631-636. **28.** Huttelmaier J, et al. *Front Immunol*. 2023;14:1196999. **29.** Kibsgaard L, et al. *Acta Derm Venereol*. 2015;95:307-311. **30.** Lee J, et al. *Front Immunol*. 2019;10.

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