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Abstract

Background Bullous pemphigoid (BP) is the most common autoimmune subepidermal blistering disease of the skin and mucous membranes. This disease typically affects the elderly and presents with itch and localized or, most frequently, generalized bullous lesions. A subset of patients only develops excoriations, prurigo-like lesions, and eczematous and/or urticarial erythematous lesions. The disease, which is significantly associated with neurological disorders, has high morbidity and severely impacts the quality of life.

Objectives and methodology The Autoimmune blistering diseases Task Force of the European Academy of Dermatology and Venereology sought to update the guidelines for the management of BP based on new clinical information, and new evidence on diagnostic tools and interventions. The recommendations are either evidence-based or rely on expert opinion. The degree of consent among all task force members was included.

Results Treatment depends on the severity of BP and patients' comorbidities. High-potency topical corticosteroids are recommended as the mainstay of treatment whenever possible. Oral prednisone at a dose of 0.5 mg/kg/day is a recommended alternative. In case of contraindications or resistance to corticosteroids, immunosuppressive therapies, such as methotrexate, azathioprine, mycophenolate mofetil or mycophenolate acid, may be recommended. The use of doxycycline and dapsone is controversial. They may be recommended, in particular, in patients with contraindications to oral corticosteroids. B-cell-depleting therapy and intravenous immunoglobulins may be considered in treatment-resistant cases. Omalizumab and dupilumab have recently shown promising results. The final version of the guideline was consented to by several patient organizations.

Conclusions The guidelines for the management of BP were updated. They summarize evidence- and expert-based recommendations useful in clinical practice.

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Conflict of interest

See attachment.

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[Correction added on 29 July 2022, after first online publication: name and affiliations for author AV Marzano have been updated.]