A systematic review on efficacy, safety, and treatment durability of intravenous immunoglobulin in autoimmune bullous dermatoses: special focus on indication and combination therapy

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## Abstract

Background: Autoimmune bullous diseases (AIBDs) are a group of rare blistering dermatoses of the mucous membrane and/or skin. The efficacy, safety, and treatment durability of intravenous immunoglobulin (IVIg) as an alternative treatment should be explored. Objectives: To systematically review the available literature regarding treatment outcomes with IVIg in AIBD patients. Methods: The predefined search strategy was incorporated into the following database, MEDLINE/PubMed, Embase, Scopus, and Web of Science on 18th July 2022. Sixty studies were enrolled using Preferred Reporting Items for Systematic Reviews and Meta-analyses guidelines. Results: The use of IVIg alone or combined with rituximab was reported in 500 patients with pemphigus, 82 patients with bullous pemphigoid, 146 patients with mucous membranes pemphigoid, and 19 patients with epidermolysis bullosa acquisita. IVIg therapy induced disease remission of 82.8% in pemphigus, 88% in bullous pemphigoid, 91.3% in mucous membranes pemphigoid, and 78.6% in epidermolysis bullosa acquisita. The combination therapy of RTX+IVIg led to disease remission of 86.7% in pemphigus, 100% in bullous pemphigoid, and 75.0% in mucous membranes pemphigoid. Among all the included patients, 37.5% experienced at least one IVIg-related side effect; the most common ones were headaches, fever/chills, and nausea/vomiting. Conclusions: The use of IVIg with or without rituximab had a favorable clinical response in patients with AIBDs. IVIg has no major influence on the normal immune system, which makes its utilization for patients with AIBDs reasonable.

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