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Efficacy, safety, and treatment durability of intravenous immunoglobulin in autoimmune blistering diseases

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Introduction & Objectives: 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.

Materials & Methods: To systematically review the available literature regarding treatment outcomes with IVIg in AIBD patients. The predefined search strategy was incorporated into the following database, MEDLINE/PubMed, Embase, Scopus, and Web of Science on 18 July 2022.

Results: Sixty studies were enrolled using Preferred Reporting Items for Systematic Reviews and Meta-analyses guidelines. 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. Disease remission with IVIg therapy and RTX + IVIg combination therapy were recorded as 82.8% and 86.7% in pemphigus, 88.0% and 100% in bullous pemphigoid, and 91.3% and 75.0% in mucous membrane pemphigoid, respectively. In epidermolysis bullosa acquisita, treatment with IVIg led to 78.6% disease remission; no data were available regarding the treatment with RTX + IVIg in this group of patients. 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. The use of IVIg with or without rituximab had a favorable clinical response in patients with AIBDs.

Conclusion: IVIg has no major influence on the normal immune system, which makes its utilization for patients with AIBDs reasonable.

Table 1** Demographic data, efficacy, and treatment durability of intravenous immunoglobulin in patients with bullous diseases

	Pemphigus	Bullous pemphigoid	Mucous membrane pemphigoid	Epidermolysis bullousa
	IVIg	RTX+IVIg	IVIg	RTX+IVIg
Patients	410	90	70	12
Clinical response, n (%)				
	Yes	290 (90.9%)	61 (100%)	33 (80.5%)
	No	29 (9.1%)	0 (0.0%)	8 (19.5%)
	NM	91	29	29
Time to clinical response, m				
	Mean	1.9	2.6	2.8
	Range	0.3-7.5	0.4-6	2-4
	NM	236	33	52
Clinical remission, n (%)				
	Yes	227 (82.8%)	78 (86.7 %)	22 (88.0%)
	No	47 (17.2%)	12 (13.3%)	3 (12.0%)
	NM	136	0	45
Treatment durability				
	Relapse, n (%)	41 (44.6%)	22 (40.8%)	10 (52.6%)
	Follow-up, m	37.2	67.8	22.7
	NM	312	27	50

IVIg, intravenous immunoglobulin; RTX, rituximab; NM, not mentioned.