

Table SII. Details of included studies

Study (year)	Definition of endpoints ^a	Scoring system for disease severity ^b
Arin et al. (4), 2005	CR: healing of mucocutaneous lesions for >2 months within 12 months after RTX PR: >50% healing; MR: <50% healing	Disease activity score as PSS (42) with slight modifications
Goh et al. (7), 2007	CR: absence of clinical disease without further treatment CRm: absence of clinical disease on medication PD: any increase in medication or extent of skin involvement CR: absence of new lesions with complete healing of old lesions for a minimum of 8 weeks regardless of the treatment used	Disease severity score adapted from PSS (42)
Marzano et al. (8), 2007	PR and MR: healing of the mucocutaneous lesions by at least 50% and less than 50%, respectively, of the body area initially involved, with the possible presence of ≤5 new lesions lasting >1 week CR: absence of new lesions with complete healing of previous lesions with or without further treatment CR: absence of lesions for at least 1 month and no treatment with CS or adjuvants or treatment with ≤5 mg of prednisone per day Long-lasting CR: no treatment with CS or adjuvant therapies for 6 months and being free of lesions PR: presence of 1–5 new oral or cutaneous blisters per week with treatment with ≤10 mg prednisone and no adjuvants	Disease activity score as PSS (42) with slight modifications
Antonucci et al. (5), 2007		BSA
Cianchini et al. (6), 2007		Ikeda severity score (ISS) (43)
Shimanovich et al. (25), 2008	Disease control: suppression of new blisters, together with the beginning of healing of the existing lesions and the Nikolsky phenomenon potentially present [AQ6] PAIA: D1–3 and then D8, 15, 22, 29 along with RTX; maintenance PAIA was stopped if healing of 80% of pre-existing lesions without fresh lesions for 1 week CR: clinical and serological remission off treatment CIr (clinical remission): healing of all lesions on further therapy PR: healing of >80% of lesions CR (score 0): complete remission and immunosuppression omitted CIr (score 1): clinical remission without lesions but immunosuppression required PR: improvement of the scores Relapse: back to baseline (score 3–4) Concomitant IA protocol: not specified Consensus (34)	Disease activity (0–4); 3–4 as baseline clinical status.
Schmidt et al. (9), 2009		Disease activity (0–4); 3–4 as baseline clinical status.
Kim et al. (11), 2011		PSS (42) with modified definitions for mild, moderate and severe (PV: 0–4, 5–8 and 9–13; PF: 0–3, 4–7 and 8–11) N/A
Kasperkiewicz et al. (10), 2011	Consensus (34) PAIA: on days 1, 2, 3 (first treatment cycle), 21, 22 and 23 (second treatment cycle), along with RTX on days 4 and 24 (1 treatment cycle). Three-day IA treatment cycles were repeated initially every 3 and then 4 weeks until skin lesions had healed by 90% Consensus (34) Consensus (34) Consensus (34) Consensus (34) Consensus (34) IA: The first cycle of IA was given within 1–6 months prior to the first administration of RTX. During a single treatment cycle, IA was administered on 4 consecutive days (2A5-fold plasma volume on each day), followed by a second IA cycle 4 weeks later. The optional third and fourth treatment IA cycles were also performed 4 weeks after the previous one Consensus (34)	
Kim et al. (22), 2011		PSS (42) with slight modifications
Kasperkiewicz et al. (28), 2012		N/A
Horváth et al. (23), 2012		N/A
Behzad et al. (26), 2012		Autoimmune Bullous Skin Disorder Intensity Score (ABSID) (44)
Reguiat et al. (13), 2012		N/A
Kasperkiewicz et al. (27), 2012		BSA and well-being (visual analogue scale)
Cianchini et al. (18), 2012		ISS (43)
Lunardon et al. (12), 2012	Consensus (34) plus CI (CR) defined as clinical improvement (complete remission on doses greater than minimal therapy) and CI (PR) defined as clinical improvement (partial remission on doses greater than minimal therapy)	N/A

Table SII. *Contd.*

Colliou et al. (35), 2013	Consensus (34)	BSA
Leshem et al. (19), 2013	Consensus (34)	N/A
Baum et al. (15), 2013	Consensus (34) but whether patients were on or off all systemic therapy at remission were not specified.	N/A
Kanwar et al. (24), 2013	Consensus (34)	ISS (43)
Kolesnik et al. (29), 2014	CR: according to consensus (34) PR: defined as presence of single small lesions	ABSIS (44)
Balighi et al. (14), 2013	PR: defined as presence of single small lesions Magdeburg treatment protocol: induction consisted of 3 initial PAIA on 3 consecutive days, followed by the first RTX infusion at day 4. Post-induction phase comprised single PAIA in weekly and later bi-weekly intervals latest up to week 31, coupled to a total of 4–6 single RTX administrations on the subsequent day. The cohort of 15 patients with autoimmune skin blistering diseases was treated with a mean of 9.3 PAIA and 4.2 RTX infusions (375 mg/m ² each) Initial clinical improvement (CI): defined as the time from the first RTX infusion to cessation of new blister formation, negative Nikolsky sign and re-epithelialization of the earlier lesions Marked CI: no new blister formation, negative Nikolsky sign and near complete re-epithelialization of the lesions parallel to prednisolone dose reduction Relapse: clinical disease progression (new blister or positive Nikolsky sign) and an inevitable increase in the dose of prednisolone for disease control after a CI. (Minor relapse was defined as <10 cutaneous or 5 mucosal lesions, with more numerous lesions characterizing major relapse.)	N/A
Cho et al. (31), 2013	Consensus (34)	PSS (42)
Heelan et al. (20), 2014	Consensus (34)	N/A
Cho et al. (16), 2013	Consensus (34)	PSS (42)
Kanwar et al. (21), 2014	Consensus (34)	ISS (43) and Saraswat's modified oral pemphigus score (SMOPS) as mild (<5), moderate (5–7) and severe (>7). Revised ISS (43)
Gregoriou et al. (17), 2014	Consensus (34)	Pemphigus Disease Area Index (PDAI)
Leshem et al. (36), 2014	Consensus (34)	

*Consensus (34) as following: "CR off therapy" was defined as the absence of new or established lesions, as well as the healing of all skin and mucosal lesions with complete epithelialization when patients were off all systemic therapy for at least 2 months.

"CR on minimal therapy" was defined as the absence of new or established lesions while patients were still receiving minimal therapy (prednisone less than or equal to 10 mg/day) and/or minimal adjuvant therapy for at least 2 months.

"PR off therapy" was defined as presence of transient new lesions that heal within 1 week without treatment and while the patient is off all systemic therapy for at least 2 months.

"PR on therapy" was presence of transient new lesions that heal within 1 week while the patient is receiving minimal therapy, including topical steroids.

"Incomplete remission" was defined as patients still receiving prednisone doses higher than 10 mg/day at the end of the study, even if they had no skin or mucosal lesions at the time of examination.

"Relapse" was defined as the occurrence of >3 new lesions/month that do not heal spontaneously within 1 week, or by the extension of established lesions after having achieved disease control.

"Treatment failure" was defined as the inability to control disease activity with full therapeutic doses of systemic treatments.

"Minimal therapy" was defined as: (i) prednisolone in a dosage of less than 10 mg daily for at least the last 2 months; and/or (ii) "steroid sparing agents" at half of the regular dosage.

^bPSS was graded as mild, moderate, or severe based on a severity score of ≤ 2 , 3–6, or ≥ 7 , respectively. (42) ISS was classified as mild (<5), moderate (5–7), or severe (>7). BSA, ABSIS and PDAI were classified as mild (<10), moderate (10–30), or severe (>30) based on authors' clinical experience and previous literature (34, 40, 45).

PAIA: protein A immunoadsorption; RTX: rituximab; CR: complete remission; PR: partial remission; MR: minimal remission; IA: immunoadsorption; N/A: no use of scoring system; PSS: pemphigus severity score; BSA: body surface area.