

Table SIII. Summary of clinical and therapeutic data of pemphigus patients from included studies and divided into high dose protocol, low dose protocol, and plus IA protocol

Study	Mean age (years)	Disease duration (years)	TDC (week) ^a	TCR on (months) ^a	CR (%) ^b	Remission duration of CR (months)	Relapse, n (%)	Major adverse event (%)	Mean follow-up time (months)
High-dose group									
Lymphoma protocol (total 2,595 mg by standardized body surface area of 1.73 m ²)									
Arintet al. (4), 2005	47.4 ± 19.4	6.8 ± 4.6	N/A	5.3	3/5 (60)	20.7	2/5 (40)	0	19.6
Goh et al. (7), 2007	54 ± 6.7	3.4 ± 2.9	N/A	5.6	3/5 (60)	10.7	0	2/5 (40), opportunistic infection	16.8
Marzano et al. (8), 2007	43.6 ± 7.6	5.2 ± 1.5	6.4	3	4/5 (80)	8.9	1/5 (20)	0	16.4
Antonucci et al. (5), 2007	30.6 ± 2.7	4.8 ± 1.6	4.4	3	5/5 (100)	9	1/5 (20)	0	12
Cianchini et al. (6), 2007	42.3 ± 10.2	3.9 ± 3.7	4	4	9/12 (75)	6.9	0	0	10.5
Schmidt et al. (9), 2009	52.4	N/A	N/A	6	23 (66.7)	9	1/3 (33.3)	0	15
Kim et al. (11), 2011	N/A	N/A	2.5	3.8	9/16 (56.3)	N/A	5/16 (31.3)	0	36
Kasperkiewicz et al. (10), 2011	47.5	3.5	N/A	3	1/2 (50)	56	1/2 (50)	0	25.4
Reguiat et al. (13), 2012	40 ± 18.9	6.9 ± 6.6	N/A	6	13/13 (100)	23.9	7/13 (53.8)	0	45.6
Lunardon et al. (12), 2012	44.9 ± 11.3	7.58 ± 6.3	N/A	N/A	4/15 (26.7)	N/A	2/15 (13.3)	0	40.8
Colliou et al. (35), 2013	53.7 ± 15.6	N/A	N/A	8.4	20/21 (95.3)	31.8	17/21 (81)	2/21 (9.5); pyelonephritis and septicemia	74
Baum et al. (15), 2013	50.9 ± 11.6	7.4 ± 6.4	N/A	5.3	13/18 (72.2)	5.3	4/18 (22.2)	0	16
Balighi et al. (14), 2013	40.5	2.9	2.4	9.1	40/40 (100)	N/A	21/41 (52.5)	4/40 (10%); lung abscess, cavernous sinus thrombosis, sepsis, SJS	12
Cho et al. (31), 2013	54.5 ± 17.1	4.1 ± 3.9	6.2 ± 3.2	4.3 ± 1.2	4/8 (50)	14.5	5/8 (62.5)	0	24.9
Gregoriou et al. (17), 2014	49 ± 16.6	5.1 ± 4.2	5	N/A	18/18 (100)	N/A	7/18 (38.9%)	0	9-48
Total lymphoma data (n=186)	46 (n=170)	5.0 (n=146)	3.6 (n=104)	6.6 (n=126)	148/186 (79.6)	18.2 (n=75)	74/186 (39.8)	8/186 (4.3)	29.1 (n=168)
RA protocol (total 2,000 mg)									
Kasperkiewicz et al. (10), 2011	42.3	1.56	N/A	3	4/4 (100)	14	3/4 (75)	0	20.5
Cianchini et al. (6), 2012	27-75	4.2	N/A	3	36/42 (86)	N/A	20/42 (47.6)	0	26.5
Lunardon et al. (12), 2012	49.2	3.3 ± 3.1	N/A	N/A	9/16 (56.3)	N/A	2/16 (12.5)	2/31 (12.5); phlegmon and osteomyelitis	23.4
Leshem et al. (19), 2013	52 (18-83)	2.1 (0-13.6)	N/A	4 (1-36)	28/45 (62.2)	N/A	5/45 (11.1)	0	18
Kanwar et al. (24), 2013	36.5 ± 12.5	1.6 ± 1.9	8	N/A	6/9 (66.7)	N/A	N/A	2/9 (22.2) sepsis	8
Heelan et al. (20), 2014	47 (17-77)	2 (0-21.3)	N/A	N/A	74/92 (80)	N/A	56/92 (61)	0	24
Kanwar et al. (21), 2014	33.2	2.1	7.1 ± 3.5	5.9	10/11 (90.9)	5.1	4/11 (36.4)	0	12
Total RA data (n=219)	42.9 (n=40)	3.3 (n=82)	7.2 (n=19)	3.8 (n=50)	167/219 (76.3)	7.6 (n=14)	90/219 (42.9)	4/219 (1.8)	21.8 (n=219)
Others (total near or ≥2,000 mg)^c									
Kim et al. (22), 2011	47.7 ± 14.3	1.7	4 (1-10)	5.0 (2.1-13.2)	11/15 (73)	N/A	0	0	18
Kasperkiewicz et al. (27), 2012	52	N/A	N/A	N/A	4/22 (18.2)	N/A	N/A	N/A	11
Cho et al. (31), 2013	54.1 ± 16.0	0	2.2	3.85	9/9 (100)	6.6	8/9 (88.9)	0	20.5
Cho et al. (16), 2013	57	3.1	5.5	3.8	2/2 (100)	24.4	2/2 (100)	0	28.3
Total high-dose data (n=453)	46.5 (n=258)	4.1 (n=254)	3.9 (n=149)	5.7 (n=185)	354/453 (78.1)	15.8 (n=100)	172/422 (40.8)	12/431 (2.8)	23.9 (n=435)

Table SIII. *Contd.*

<i>Low-dose group (total <1,500 mg)</i>												
Kasperkiewicz et al. (10), 2011	50	0.33	N/A	3	1/1 (100)	9	0	0	0	0	12	
Kim et al. (22), 2011	48.4 ± 10.1	3.5 ± 2.1	3 (1–11)	14.4 (3.1–29.2)	5/12 (42)	N/A	8/12 (67)	0	0	0	11.5	
Horváth et al. (23), 2012	55.1 ± 15.1	6.1 ± 3.2	9.8	11.5	8/15 (53.3)	7.1	6/15 (40)	1/15 (6.7)	sepsis	0	21.3	
Cho et al. (16), 2013	51.5 ± 13.2	4.4 ± 3.0	3.9	3.5	9/13 (69.2)	15.3	3/13 (23.1)	0	0	0	18	
Kanwar et al. (21), 2014	33.5	1.3	7.4 ± 4.5	4.2	11/11 (100)	5.5	7/11 (63.6)	0	0	0	12	
Total low-dose data (n = 52)	48.0 (n = 52)	4.1 (52)	6 (n = 51)	6.1 (n = 29)	34/52 (65.4)	9.1 (n = 29)	24/52 (46.2)	1/52 (1.9)			16.6 (n = 52)	
<i>Rituximab (high dose) plus immunoadsorption protocol</i>												
Shimanovich et al. (25), 2008	56.9 ± 16.9	2.9 ± 2.3	-2 ^d	3.75	3/7 (42.9)	16.3	3/7 (42.9)	3/7 (42.9); PCP, staphylococcal bacteremia and PE			23.3	
Schmidt et al. (9), 2009	52.4	N/A	N/A	5	3/8 (37.5)	10	4/8 (50)	0	0	0	15	
Kasperkiewicz et al. (10), 2011	53.3	2.7	N/A	3.3	3/3 (100)	9.3	3/3 (100)	0	0	0	29	
Kasperkiewicz et al. (28), 2012	54.3 ± 12.7	3.0	-1.3 ^d	8.2	22/23 (95.7)	17.7	6/23 (26)	2/23 (9); staphylococcal bacteremia and extensive herpes simplex infection			29	
Behzad et al. (26), 2012	52.1 ± 9.8	5.4 ± 2.6	N/A	3.3	8/10 (80)	7.3	1/10 (10)	0	0	0	10.8	
Kasperkiewicz et al. (27), 2012	52	N/A	N/A	N/A	4/14 (29)	N/A	N/A	N/A			11	
Kolesnik et al. (29), 2014	59.5 ± 13.3	4.0 ± 6.7	-1 ^d	4.7	7/8 (87.5)	20.6	1/8 (12.5)	0	0	0	20.5	
R+IA total data (n = 73)	54.1 (n = 73)	3.6 (n = 51)	N/A	6.1 (n = 46)	50/73 (68.5)	15.2 (n = 46)	18/59 (30.5)	5/73 (8.5)			21.1 (n = 73)	
Overall total data (n = 578)	48.2 (n = 383)	4.1 (n = 357)	4.4 (n = 200)	5.8 (n = 262)	438/578 (75.8)	14.5 (n = 175)	214/533 (40.2)	18/542 (3.3)			22.9 (n = 560)	

^aStarted from the end of rituximab (RTX) treatment. ^bComplete remission according to the definition of the original study (Table S1). ^cThe group classified as "Others" included patients receiving high-dose regimens other than RA or lymphoma protocols and those using high-dose RTX without detailed information in Kasperkiewicz et al. (17), 2012. ^dOnset immediately after starting IA and before completion of the RTX regimen.

Data expressed as mean ± standard deviation (if available) or percentage.

Comparisons by analysis of variance (ANOVA) or χ^2 test.

CR: complete remission; TDC: time to disease control; TCRon: time to complete remission on therapy; IA: immunoadsorption; RA: rheumatoid arthritis; SJS: Stevens–Johnson syndrome; PCP: pneumocystosis carinii pneumonia; PE: pulmonary embolism; N/A: not applicable.