

Table S1: Patient demographics and baseline disease characteristics of patients receiving bortezomib.

Pat. ID	Sex/ Age/ race	Clinical manifestations		Treatment		SLED AI before Bz	Bortezomib treatment		SLEDAI after last Bz cycle	Therapy 6 months after last Bz cycle	SLEDAI 6 months after last Bz cycle
		History	Present at baseline	History	Present at baseline		Start Date (mm/yyyy)	No. of Bz cycles			
BE #1	m/24/c	Nephritis (WHO IV), CNS, serositis, musculoskeletal, mucocutaneous, hematologic, myositis	Nephritis, arthritis, myositis, rash, alopecia	CYC, AZA, MTX, HQL, ASCT (-6y), RTX (2x1g, -13 mo)	CYC (12x1g monthly, -158d), HQL, 20mg Pred	20	05/2009	4	4	CYC, HQL 10mg Pred	8
BE #2	f/33/c	Vasculitis, musculoskeletal, mucocutaneous, hematologic, myositis	Vasculitis, arthritis, rash, alopecia, hematologic	CYC, MMF, AZA, HQL, IA, RTX (2x1g, -13 mo)	CYC (6x1g monthly, -180d), HQL, 50mg Pred	20	05/2010	2	7	MTX, HQL 15mg Pred	7
BE #3	f/42/c	Polyserositis, musculoskeletal, mucocutaneous, hematologic	Arthritis, serositis, alopecia, hematologic	CYC, AZA, HQL	CYC (6x500mg monthly, -38d), HQL, 5mg Pred	13	05/2010	4	2	CYC, HQL, 5mg Pred	4
BE #4	f/30/c	Nephritis (WHO IV), serositis, musculoskeletal, mucocutaneous,	Arthritis, serositis, alopecia	CYC, MMF, AZA, HQL, IA RTX (2x1g, -20mo)	CYC (6x1g monthly, -179d), 20mg Pred	10	12/2010	4	2	HQL, AZA, 7.5mg Pred	4
BE #5	m/26/b	Nephritis (WHO IV), musculoskeletal, mucocutaneous	Nephritis, rash, nephrotic syndrome	CYC, AZA, HQL	CYC (6x1g monthly, -32d), HQL, RTX (1x1g), 10mg Pred	10	08/2011	1	4	MMF, RTX, HQL, 7.5mg Pred	2
BE #6	f/35/c	Arthritis, musculoskeletal, mucocutaneous	Arthritis, rash, oral ulcer, alopecia	AZA, MTX, LEF, HQL	AZA (100mg/d, -24d), HQL, 10mg Pred	14	12/2011	1	4	AZA, HQL, BEL, 10mg Pred	6
BE #7	m/35/c	Nephritis (WHO IV), CNS, serositis, musculoskeletal, mucocutaneous, APS, hematologic,	Arthritis, rash, terminal renal failure, hematologic	CYC, AZA, MTX, HQL, IVIG, CsA, TAC, RTX (2x1g - 24mo)	10mg Pred	10	07/2012	1	4	BEL, 10mg Pred	4
BE #8	f/40/c	CNS, serositis, musculoskeletal, mucocutaneous, APS, hematologic	Arthritis, serositis, alopecia, hematologic	CYC, AZA, MTX, HQL, MMF, IVIG	10mg Pred	13	11/2012	2	4	BEL, 7.5mg Pred	5
ER #1	f/30/c	Nephritis (WHO IV), serositis, musculoskeletal, mucocutaneous, hematologic	Arthritis, serositis, rash, oral ulcer, terminal renal failure	MTX, AZA, CsA, CQN, TAC, MMF, CYC, RTX (2x1g, -7mo),	CYC (1g monthly, -30d), 60mg Pred	18	05/2008	2 ¹	7	HQL, 5mg Pred	8
ER #2	f/25/c	Nephritis (WHO IV), CNS, musculoskeletal, mucocutaneous, hematologic	Nephritis, arthritis, rash, alopecia	MMF, AZA, MTX, CQN, HQL, LEF, RTX (3x1g, -63d),	CYC (1g monthly, -28d), 20mg Pred	21	12/2008	2 ¹	8	TAC, 5mg Pred	4
ER #3	f/34/c	Nephritis (WHO IV), CNS, vasculitis, musculoskeletal, mucocutaneous, hematologic	Nephritis, arthritis	CQN, CYC, MMF, RTX (2x1g, -17mo)	CYC (1g monthly, -21d), 50mg Pred	14	12/2008	3 ¹	6	MMF, HQL 5mg Pred	2
KO #1	m/29/c	Nephritis (WHO IV), CNS, vasculitis, serositis, musculoskeletal, fever, hematologic	Nephritis, arthritis, myositis, hematologic	MTX, MMF, AZA, CsA, HQL, TAC, RTX (2x1g, -36mo), ASCT (-2y)	MMF (2g/d, -8d), 100mg Pred	18	02/2009	4	8	MMF, 10mg Pred	4

APS = antiphospholipid syndrome, ASCT = autologous stem cell transplantation, AZA = azathioprine, Bz = bortezomib, BEL = belimumab, CQN = chloroquine, CNS = central nervous system, CYC = cyclophosphamide, CsA = ciclosporine A, HQL = hydroxychloroquine, IA = immunoadsorption, IVIG = intravenous immunoglobulins, LEF = leflunomide, MMF = mycophenolate mofetil, MTX = methotrexate, Pred = prednisolone, RTX = rituximab, TAC = tacrolimus, race: c = caucasian, b = black, time baseline treatment was discontinued before bortezomib as depicted in brackets: y = years, mo = months, d = days.

¹ Bortezomib cycle consisted of treatment on day 1, 4, 8

Table S2: Patient demographics and baseline disease characteristics of patients receiving bortezomib without coadministration of dexamethasone.

Pat. ID	Sex/ Age/ race	Clinical manifestations		Treatment		SLED AI before Bz	Bortezomib treatment		SLEDAI after last Bz cycle	Therapy 6 months after last Bz cycle	SLEDAI 6 months after last Bz cycle
		History	Present at baseline	History	Present at baseline		Start Date (mm/yyyy)	No. of Bz cycles			
ER #4	f/34/c	Nephritis (WHO), serositis, musculoskeletal, mucocutaneous, hematologic	Nephritis, hematologic	AZA, MMF, CYC, CsA, HQL, RTX (2x1g, -297d), TOC (-120d)	TAC, MMF, 7.5 mg Pred	9	09/2009	4	8	RTX, MMF, 5mg Pred	4
ER #5	f/30/c	Serositis, musculoskeletal, mucocutaneous, hematologic	Arthritis, fever	MTX, AZA, CQN, MMF, CYC, ETA, IA, RTX 2x1g, -159d),	CYC (2x800mg, -96d) 50mg Pred	8	10/2009	6	2	RTX, MMF, 5mg Pred	2
HE #1	f/22/c	Nephritis (WHO IV), CNS, vasculitis, APS, serositis, hematologic	Nephritis, CNS, vasculitis, serositis	CYC, AZA, RTX (2x1g, -17d)	CYC 1g monthly, -30d 500mg Pred	33	12/2011	4 ¹	12	MMF, HQL, 5mg Pred	4
FR #1	f/42/c	Nephritis (WHO IV), CNS, serositis	Nephritis with terminal renal failure	AZA, CYC, MMF, CsA	CYC (6x1g, -143d), PP, BEL (-12d) 5mg Pred	22	05/2012	3	14	5mg Pred	17

APS = antiphospholipid syndrome, AZA = azathioprine, BEL = belimumab, CQN = chloroquine, CNS = central nervous system, CYC = cyclophosphamide, CsA = ciclosporine A, ETA = etanercept, HQL = hydroxychloroquine, IA = immunoabsorption, MMF = mycophenolate mofetil, MTX = methotrexate, PP = plasmapheresis, Pred = prednisolone, RTX = rituximab, TAC = tacrolimus, TOC = tocilizumab, race: c = Caucasian, time baseline treatment was discontinued before bortezomib as depicted in brackets: y = years, mo = months, d = days.

¹ Bortezomib cycle consisted of treatment on day 1, 4, 8

Table S3: List of adverse events in SLE patients (n=12) during bortezomib treatment.

Body System / ADR term	Number of adverse events	Severity Grade	Severe adverse event	Outcome	Adverse event leading to discontinuation of Bortezomib
Allergy/Immunology	4				
Fever	2	mild	2	resolved	2
Allergic skin reaction	2	mild	0	resolved	2
Gastrointestinal	4				
Nausea	3	mild	0	resolved	0
Diarrhea	1	mild	0	resolved	0
Infection	3				
Herpes zoster	1	moderate	1	resolved	0
Common cold	1	mild	0	resolved	0
Urinary tract infection	1	mild	1	resolved	1
Musculoskeletal/Soft tissue	1				
Myalgia	1	mild	0	resolved	0
Metabolic/Laboratory	2				
Thrombocytopenia	2	mild		resolved	
Neurology	5				
Polyneuropathy	2	mild	0	resolved	2
Headache	3	mild	0	resolved	0
Total	19		4/19		7/19