Human neutrophil peptides and complement factor Bb in pathogenesis of acquired thrombotic thrombocytopenic purpura

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Table 1s. Demographic, clinical, and laboratory data, treatment provided and long-term outcome of 52 patients with acquired thrombotic thrombocytopenic purpura

UTI-urinary tract infection; W-white; WBC-white blood cell count; *rF-WF73 assay developed in house; **FRETS-VWF73 was performed in the reference laborator for activity and inhibitor titer.

erial#	Patient Sample ID	Age	Sex	Race	CNS	Disease activity	Total episodes	s Comorbidity	WBC (x10 ³ /μl)	Neutrophil %	ANC/μΙ	Hct (%)	Platelets (x10³/µl)	LDH (U/L)	Cr (mg/dL)	ADAMTS13 activity (%)*	ADAMTS13 activity (%)**	Inhibitors (BU/ml)**	Anti-A13 IgG (U/ml)	HNP1-3 (ng/ml)	iC3b (µg/ml)	sC5b-9 (µg/ml)	C4d (µg/ml)	Bb (µg/ml)	TPE	Corticoste roids	Rituximab	Outcomes	F/U (D/
1	61	37	F	W	Yes	Initial	1	NONE	21.5	86	18447	21	27.9	952	1.6	0.0	<5	>8	14624.7	26.5	23.4	3.1	1.97	4.8	n/a	n/a	n/a	Remission	34 M
2	62	59	F	AA	No	Initial	1	CAD, HTN, CHF, PVD, HLD	18.2	87	15817	24	15.5	1968	2.5	8.1	9	< 0.4	1078.0	40.0	12.7	1.4	1.70	4.0	Yes	n/a	n/a	Remission	9 D
1	1	39	M	W	Yes	Initial	1	None	12.7	79	10033	23	17.8	941	1.3	3.8	<5	4.8	449.9	40.6	8.9	0.6	1.7	1.5	Yes	Yes	Yes	Remission	48 M
1	64	49	M	W	No	Initial	2	NONE	9.5	69	6555	28	8.9	892	2.3	1.9	<5	1.2	2939.0	31.7	13.6	1.5	3.38	3.5	Yes	Yes	No	Remission	29 D
5	4	55	M	AA	Yes	Initial	2	Nethrolithiasis	8.0	46	3671	17	8.2	1272	1.3	2.2	<5	8.0	5111.1	48.4	13.3	1.7	2.5	5.5	Yes	Yes	Yes	Remission	27 D
6	6	22	F	AA	Yes	Initial	1	NONE	21.3	50	10660	26	14	862	0.7	2.8	<5	4.8	986.5	101.9	8.5	0.7	1.9	1.3	Yes	Yes	Yes	Remission	22 D
7	67	47	F	AA	No	Initial	1	HTN, DM2, GERD	20.8	79	16432	25	108	226	0.9	2.6	<5	6.8	8551.2	17.9	3.8	0.6	1.5	0.9	No	Yes	Yes	Remission	96 N
8	72	58	M	AA W	Yes	Initial	1	AMI, CKD, HPN	9.3	79	7347	29	36.4	393	1.2	0.0	<5	0.9	2880.9	89.7	8.5	0.5	5.33	3.5	Yes	No	No	Died	13 🗅
10	75	23	M	AA	Yes	Initial	2	None	7.7 8.9	64 64	4928 5696	30 21	14.4	352	0.9 1.5	0.1	<5	1.8	13587.4	15.8	6.4	1.2	1.1 4.9	1.1 3.7	Yes	Yes	Yes	Remission	34 N 48 N
11	75	26	M	W	No No	Initial Initial	1	NONE NONE	4.6	43	1978	25	8.2	1273 565	1.5	0.0 1.1	<5 <5	1.3	3115.9 3265.4	35.6 29.3	19.9 21.0	2.3	9.86	1.6	Yes Yes	Yes Yes	No	Remission Remission	52 N
12	76	49	F	AA	No No	Replased	2	N/A	21.7	63.0	13671	18	19.2	3116	1.1	6.0	<5 <5	0.8	2811.7	29.3	30.8	3.6	3.77	8.0	n/a	n/a	n/a	Died	1 D
13	10	26		AA	No	Initial	4	NONE	4.5	53	2385	28	42	501	0.7	0.5	<5	2.8	7228.4	19.0	14.8	0.9	2.9	1.5	Yes	Yes	Yes	Remission	60 N
14	77	54	E	W	Yes	Initial	1	HTN, Gout, DP, TIA	13.3	88	11704	25	15.3	565	1.7	4.0	8	0.5	7634.3	21.7	7.7	0.9	1.5	2.6	Yes	Yes	Yes	Died	8 D
15	17	37	E	AA	Yes	Initial	1	Morbid Obesity	11.4	73	8322	16	14	733	0.9	0.0	<5	0.9	9200.6	21.7	5.9	0.8	1.6	1.7	Yes	Yes	Yes	Remission	21 N
16	20	61	E	AA	Yes	Initial	1	HTN, HPTy, Neuropathy, DM, RA, DP, SZ	5.2	n/a	n/a	19	6	446	1.8	2.3	<5	0.8	6338.1	96.0	14.3	1.9	2.9	9.6	Yes	Yes	No	Remission	32 N
7	82	33	M	AA	No	Excerbated	1	HIV on HAART	7.3	51	3743	20	16.8	806	1.0	2.3	<5	>8	13306.3	12.9	27.5	3.2	6.01	2.4	Yes	No	No	Remission	13 [
8	22	26	F	AA	Yes	Initial	2	Glomerunephritis	11.1	62	6882	22	9.7	532	1.4	4.3	<5	<0.4	4156.6	30.4	11.2	1.5	2.0	3.8	Yes	Yes	No	Remission	6 M
9	83	31	M	W	Nn	Initial	1	pneumonia and empyema	16.0	71	11360	26	14.7	2134	0.1	0.0	<5	1,1	13055.9	126.0	22.8	2.5	3.15	6.7	Yes	No	No	Remission	23 E
0	32	48	F	W	No	Relapsed	4	Obesity, UL	6.3	50	3170	31	36.4	912	0.8	0.7	<5	1.7	3213.3	20.0	12.7	0.9	4.0	3.0	Yes	Yes	Yes	Remission	8 M
1	88	56	м	AA	Yes	Refractory	1	Peumonia, AKI, MI, BT, ENP.	17.3	70	12110	16	16.8	1641	8.5	0.9	<5	8	12072.4	114.0	23.6	2.3	3.0	3.6	Yes	Yes	Yes	Died	30 [
2	24	78	M	w	Yes	Initial	1	HTN, HLD, gout	13.8	83	11454	27	21.6	694	1.2	3.3	<5	1.1	5223.8	26.7	9.5	0.8	2.0	1.5	Yes	Yes	Yes	Remission	41M
3	26	44	M	AA	No	Relapsed	4	HTN	9.2	72	6624	40	15	460	2.6	0.3	<5	1,6	2821.9	57.1	9.9	1.3	1.6	3.2	Yes	Yes	Yes	Remission	7 M
1	89	50	M	AA	n/a	Initial	1	HTN, DM2	12.1	76	9196	23	6.8	1033	1.5	1.5	<5	1.9	1541.1	76.1	33.3	3.3	14.0	3.4	Yes	Yes	No	Remission	631
5	28	56	F	AA	No	Initial	2	HTN, Obesity	12.9	67	8643	21	42	690	1.1	4.2	<5	<0.4	813.9	27.7	8.8	0.8	1.7	1.8	Yes	Yes	No	Remission	11 /
6	91	34	M	AA	Yes	Relapsed	4	HTN, CVA, SZ	17.7	79	13983	26	8.8	1539	1.3	0.0	<5	3.2	5022.2	51.2	29.8	2.9	5.6	3.0	Yes	Yes	Yes	Remission	421
27	95	84	F	AA	Yes	Initial	2	HTN, DM, DP	10.3	61	6283	26	20.3	277	1.6	0.9	<5	3.2	4368.5	36.2	10.1	1.1	3.2	4.1	Yes	Yes	Yes	Remission	24 [
28	29	32	M	AA	Yes	Initial	2	None	10.1	56	5656	20	4.7	1849	2.5	1.1	13	< 0.4	3292.7	33.5	10.7	2.2	1.9	4.5	Yes	Yes	Yes	Remission	11 7
29	97	52	F	AA	No	Initial	1	HTN, TIA	21.1	78	16458	27	48.8	848	0.7	2.0	<5	6	7494.2	12.8	9.3	0.7	3.0	1.4	Yes	Yes	No	Remission	8 D
30	99	69	F	W	No	Relapsed	4	Ob, HTN, liver Ca	13.1	70	9170	26	10.5	308	1.0	0.4	<5	3.2	4153.2	29.5	20.6	1.8	5.2	4.9	Yes	yes	Yes	Remission	471
31	101	54	F	w	Yes	Initial	1	HTN	4.9	75	3675	23	7.4	7332	3.6	0.0	<5	3.2	954.1	33.3	17.3	6.2	1.4	6.4	Yes	No	No	Died	1 D
12	56	52	M	AA	Yes	Replased	2	HTN, S.	14.5	90	13050	25	8.2	757	1.8	2.9	<5	>8.0	8777.6	46.9	4.0	0.8	1.2	1.0	Yes	Yes	Yes	Remission	2 M
13	36	47	F	AA	Yes	Initial	1	HTH, Bacterimia	24.5	78	19110	16	15	1339	3.4	0.0	<5	7.2	11102.6	28.4	8.4	0.7	1.9	2.2	Yes	Yes	Yes	Died	10 E
14	39	29	M	AA	Yes	Relapsed	5	HTN, SA, MRSA	12.9	79	10191	26	15.8	326	1.6	2.4	<5	6.6	4643.7	115.9	13.6	0.9	2.7	5.1	Yes	Yes	Yes	Remission	17 N
5	102	55	М	w	Yes	Initial	1	N/A	9.5	82	7790	25	7.6	551	1.4	0.0	<5	1.30	5394.7	23.9	17.3	1.7	1.2	1.5	Yes	No	Yes	Died	5 D
16	103	30	F	AA	No	Initial	1	Ob, SLE, SZ and asthsma	12.9	54	6966	14	7.9	1059	0.8	0.0	<5	1.1	4754.7	25.8	24.2	1.6	2.55	3.3	Yes	No	No	Remission	901
37	58	39	F	His	Yes	Initial	2	UTI	10.4	67	6968	25	5.4	1193	1.6	5.5	<5	1.9	5516.9	14.7	4.2	8.0	1.24	1.0	Yes	Yes	Yes	Remission	5 M
18	105	46	M	AA	No	Relapsed	2	PVD, HTN, HLD	9.3	76	7091	26	11.9	1042	0.7	0.6	<5	1.3	7096.0	44.7	58.6	7.4	4.31	2.6	No	No	No	Remission	62 N
39	106	49	F	AA	No	Initial	Multiple		14.4	63	9059	24	19.4	865	1.3	0.0	<5	<0.4	653.3	15.8	25.5	1.0	9.13	2.3	n/a	n/a	n/a	Died	44 [
10	54	48	M	AA	Yes	Initial	1	HTN, DM	6.9	74	5106	26	15	2494	1.4	4.8	<5	0.9	2064.0	126.9	14.3	1.7	2.6	5.3	Yes	No	No	Remission	10 E
41	107	51	F	AA	No	Initial	1	HIV, Ob, Sepsis, pneumonia	7.5	59	4431	22	15.5	2076	1.7	0.0	<5	<0.4	5471.6	24.4	27.8	2.7	4.18	3.8	Yes	N/A	n/a	Remission	56 N
12	109	58	М	AA	Yes	Initial	1	HTN, DM, CKD	12.4	65	8060	20	18.7	1343	3.4	5.7	<5	1.8	1729.1	47.1	7.7	2.5	1.8	2.6	Yes	Yes	No	Died	15 [
13	110	47	F	W	No	Initial	2	MCTD	18.2	74	13461	27	77.9	273	0.7	6.0	<5	0.5	13149.9	46.6	9.2	1.6	1.86	0.9	Yes	Yes	Yes	Remission	84 1
4 5	117	42	F	AA	n/a	Initial	1	Ads, Ob	10.8	69	7424	28	10.2	1383	1.5	0.0	<5	1.8	10892.4	47.0	32.5	1.2	6.11	2.9	Yes	Yes	Yes	Remission	421
	118	30	F	AA	No	Initial	1	HTN, DM, Asthma, GERD	15.1	65	9789	24	9.1	688	0.8	0.0	<5	1.3	10202.0	70.8	16.5	0.8	3.13	1.6	Yes	Yes	Yes	Remission	19
ô	44	34	M	AA	No	Initial	2	HTN, Ob	10.4	72	7488	32	8.9	973	1.2	1.3	<5	0.9	2063.2	24.6	16.2	1.0	2.0	2.5	Yes	Yes	No	Remission	39
7	45	34	M	AA	Yes	Relapsed	4	HTN, DM2	8.5	80	6800	16	14.1	2401	2.2	6.0	<5	6.4	8224.8	38.0	11.7	1.7	5.5	4.4	Yes	Yes	Yes	Remission	37
18	49	33	М	AA	No	Initial	2	NONE	9.5	66	6270	21	12	424	1.8	2.4	<5	4	7571.1	21.8	13.5	0.8	2.4	1.6	Yes	Yes	No	Remission	5 N
19	47	52	-	w	Yes	Relapsed	2	HTN, multiple stroke, MRSA	16.1	71	11431	29	7.9	787	0.9	1.9	<5	3.2	6253.5	26.3	7.5	1.0	3.9	1.5	Yes	Yes	Yes	Died	13
50	60	24	F	AA	No	Initial	2	NONE	5.8	67	3886	21	14.1	1575	0.8	0.6	<5	3.2	6394.5	37.0	7.1	0.7	2.0	1.8	Yes	Yes	Yes	Remission	5 N
51	120	34	M	AA	Yes	Relapsed	2	NONE	15.7	66	10362	34	44.6	435	1.3	0.0	<5	8	9433.8	26.0	15.2	1.1	3.6	1.9	Yes	Yes	Yes	Remission	411
52 african arr	50	43	M	AA	No	Initial	1	HTN, S	20.2	79	15958	28	13	1006	0.9	5.0	<5	1.9	4385.6	32.9	6.7	0.7	1.4	1.0	Yes	Yes	Yes	Remission	16 D

ADAMTS13 ADAMTS13 Inhibitors Anti-A13 InG HNP1-3 iC3h

Corticoste

HAAT-highly active antiretroviral therapy; Hct-hematocnit; His-hispanic, HIV-human immunodeficiency virus; HLD-hyperipidemia; HPTy- hyperparathyroidism; HTN-hypertension; LCa-lung cancer stage IV; LDH-lactate dehydrogenase; HNP-human neutrophil peptide; M-male; MA-macrocytic anemia, MCTD- mixed connective-lissue disease; n/a-not determined or available MRSA-methicillin-resistant staphylococcus aureus; NA-not applicable; ND-not determined or unknown; NP-neuropathy, Ob-Obesity; PVD-peripheral vascular disease; RA-rheumatoid arthritis; RF-renal failure; S-substance abuse; SA-sleep apnea; SLE-systemic lupus erythematosus; SZ-seizure; TPE-therapeutic plasma exchange; UL-uterine leiomyoma;