

Supplemental Materials:

Supplemental Figures 1 – 7

Supplemental Figure 1: Sorting strategy for new emigrant/transitional and mature naïve B cells before and after gene therapy.

Supplemental Figure 2: ~~New emigrant/transitional and mature naïve B cells B cell IgH repertoire abnormalities in WAS patients are corrected after gene therapy.~~ IgH repertoire characteristics of new emigrant/transitional and mature naïve B cells from WAS patients, before and after gene therapy.

Supplemental Figure 3: Increased BCR-induced B cell activation in WAS patients.

Supplemental Figure 4: Increased IgM cell surface expression on mature naïve B cells from WAS patients.

Supplemental Figure 5: ~~The peripheral B cell tolerance checkpoint is defective in WAS patients.~~ Frequency of polyreactive antibodies among mature naïve B cells from WAS patients.

Supplemental Figure 6: Altered Treg phenotype in WAS patients.

Supplemental Figure 7: ~~Mature naïve B cells from WAS patients post-GT express normal frequencies of polyreactive antibodies.~~ Frequency of polyreactive antibodies among mature naïve B cells from WAS patients after gene therapy.

Supplemental Tables 1- 19

Table S1: Description of WAS patients treated by LV-mediated HSC-GT

Table S2: Repertoire and reactivity of antibodies from new emigrant B cells of HD30

Table S3: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 1 before GT

Table S4: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 2 before GT

Table S5: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 3 before GT

Table S6: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 4 before GT

Table S7: Repertoire and reactivity of antibodies from mature naïve B cells of HD30

Table S8: Repertoire and reactivity of antibodies from mature naïve B cells of WAS

patient 1 before GT

Table S9: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 2 before GT

Table S10: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 3 before GT

Table S11: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 4 before GT

Table S12: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 1 after GT

Table S13: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 2 after GT

Table S14: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 3 after GT

Table S15: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 4 after GT

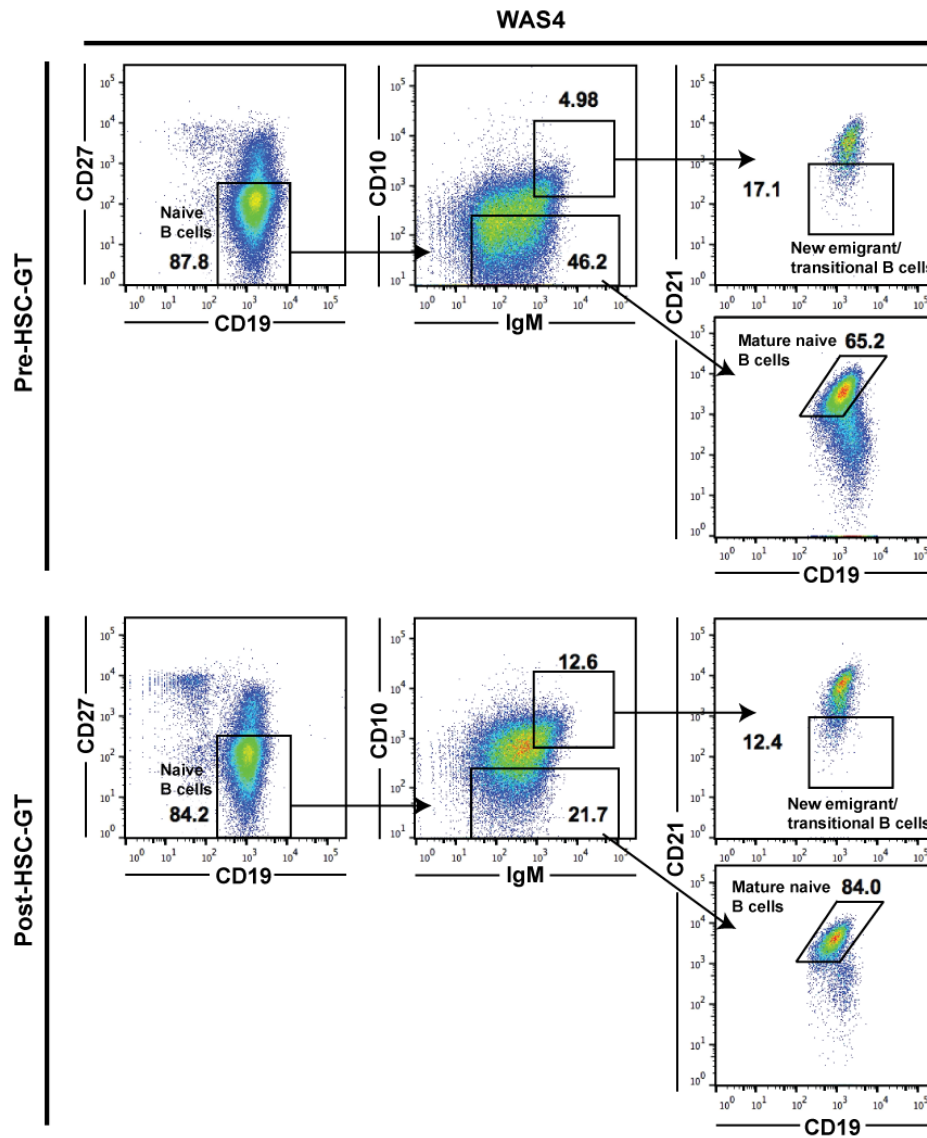
Table S16: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 1 after GT

Table S17: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 2 after GT

Table S18: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 3 after GT

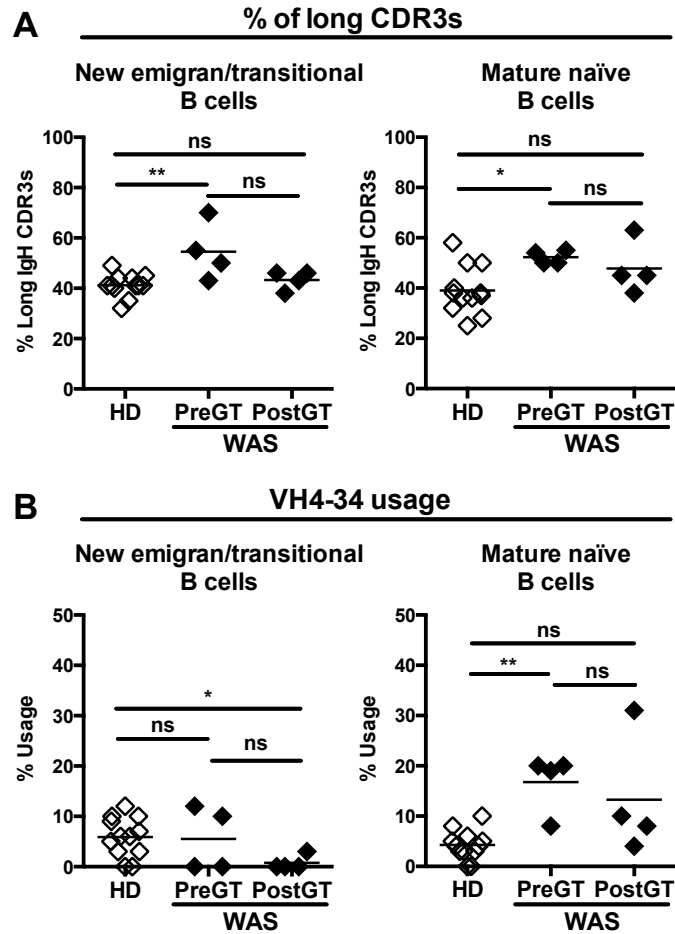
Table S19: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 4 after GT

Supplemental figures



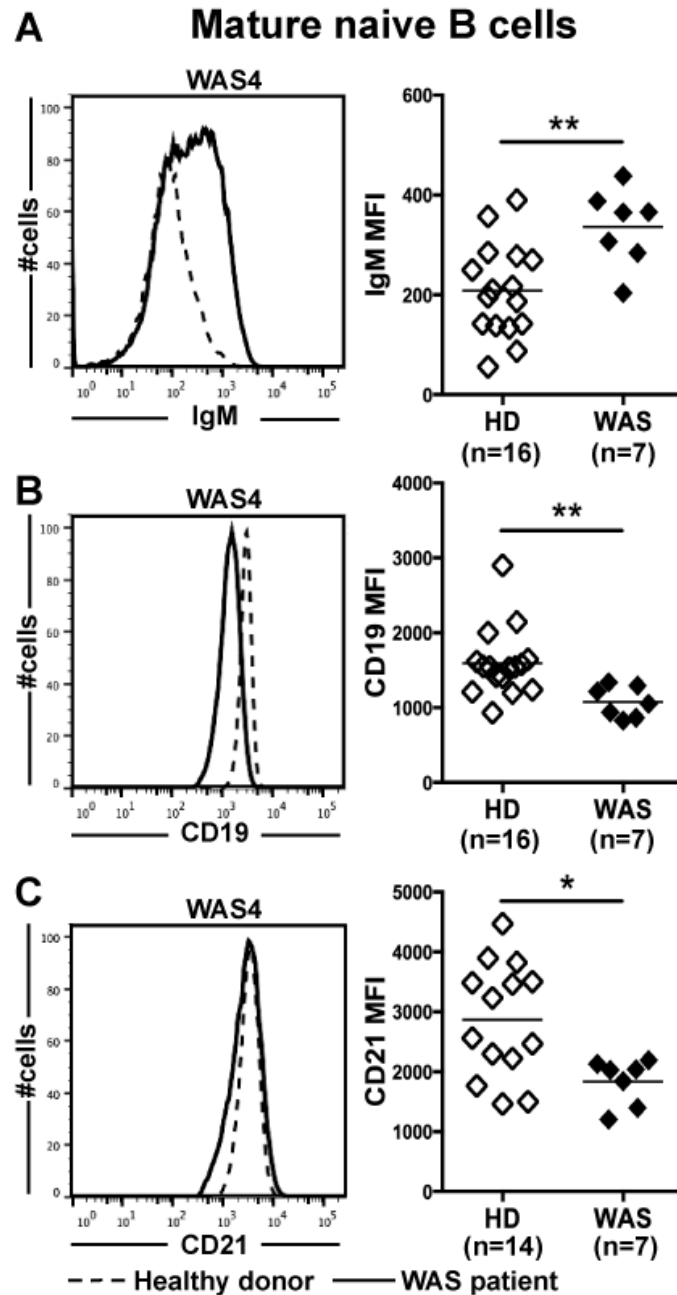
Supplemental Figure 1

Sorting strategy for new emigrant/transitional and mature naive B cells before and after gene therapy. CD19⁺CD27⁻ naive B cells were first gated and further divided into new emigrant/transitional (IgM^{hi}CD10^{hi}CD21^{low}) and mature naive (IgM⁺CD10⁻CD21⁺) B cell subsets.



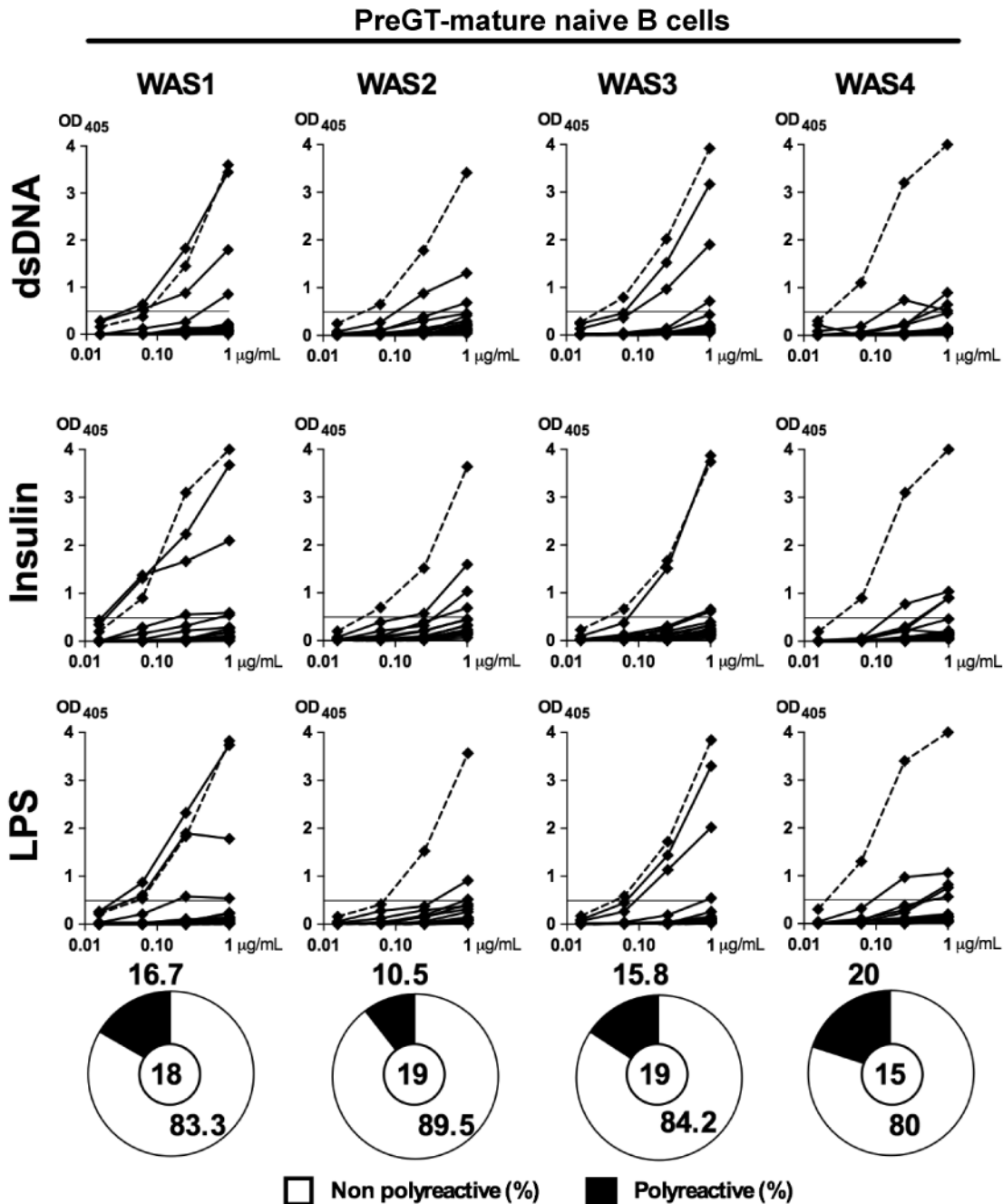
Supplemental Figure 2

~~New emigrant/transitional and mature naive B cells B cell IgH repertoire abnormalities in WAS patients are corrected after gene therapy.~~ IgH repertoire characteristics of new emigrant/transitional and mature naive B cells from WAS patients, before and after gene therapy. (A) Frequencies of long CDR3s (>14 aa) and (B) IgH CDR3s containing two or more positively charged aa in new emigrant/transitional and mature naive B cells are represented for 11 healthy control subjects and four WAS patients before and after gene therapy. (C) VH4-34 gene-usage frequencies in new emigrant/transitional and mature naive B cells. Each diamond represents an individual; the average is shown with a bar. Mann-Whitney test was used to assess statistical significance for comparisons between patients and HDs and the paired t tests for comparisons before and after gene therapy treatment. Differences reaching statistical significance are indicated. * $p < 0.01$, ** $p < 0.001$, ns: non significant.



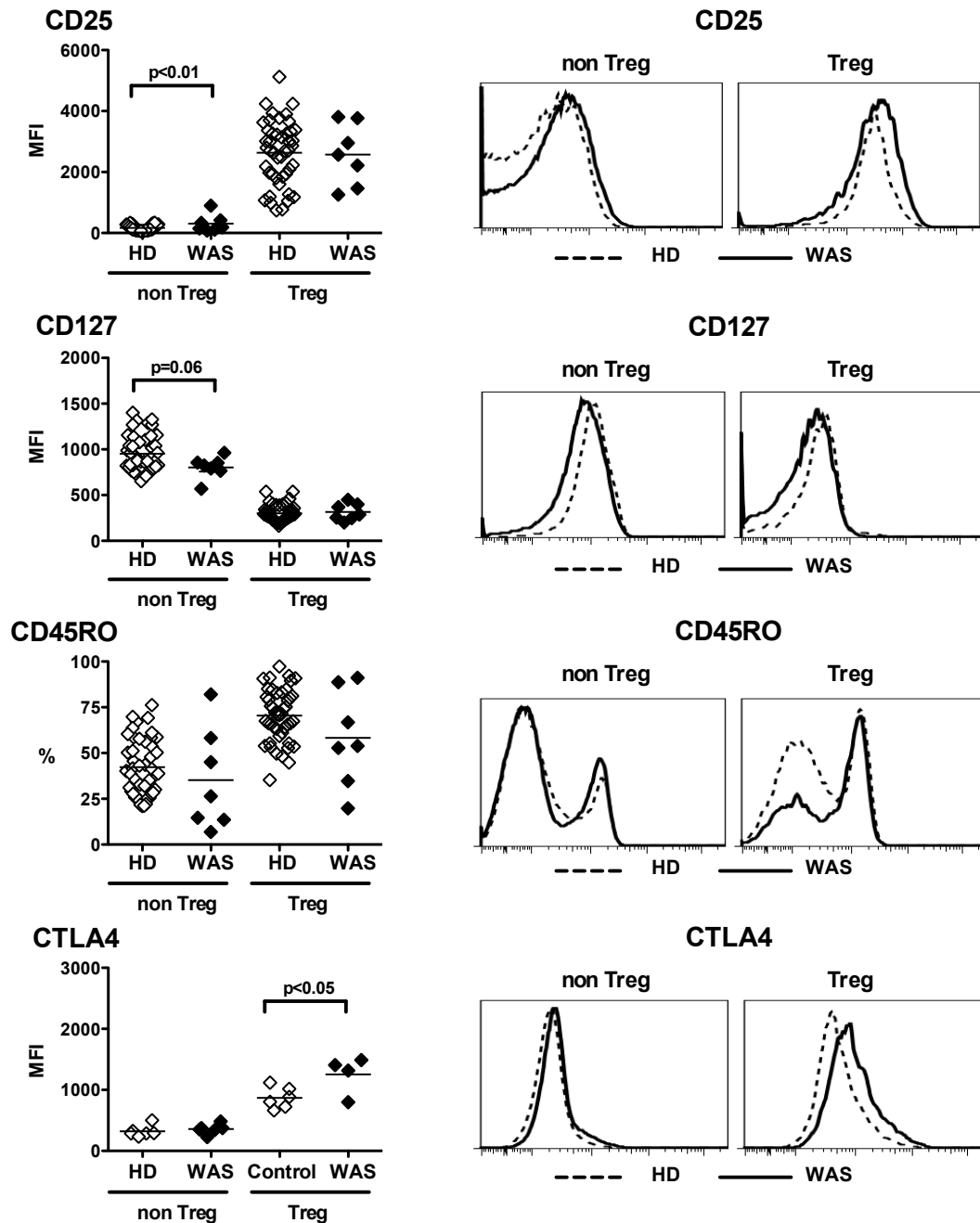
Supplemental Figure 4

Increased IgM cell surface expression on mature naïve B cells from WAS patients. Expression levels of (A) IgM, (B) CD19 and (C) CD21 on CD19⁺CD27⁻CD10⁻CD21⁺ mature naïve B cells from HD controls and WAS patients are shown as mean fluorescence intensities (MFI). Representative flow cytometry histograms of three HD control and three WAS patient are shown on the left. Differences were analyzed for statistical significance using Mann-Whitney test. *p < 0.01, **p < 0.001.



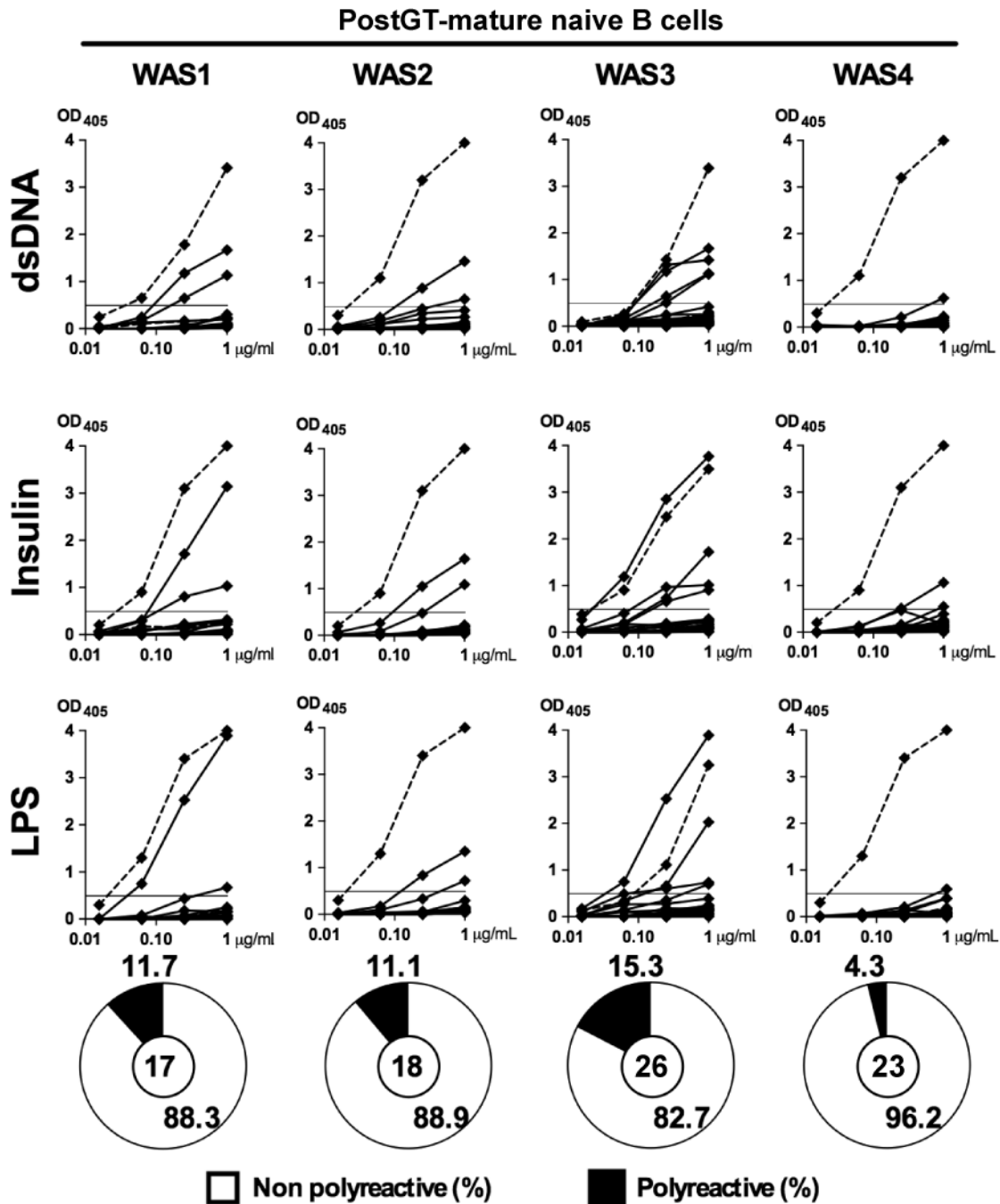
Supplemental Figure 5

The peripheral B-cell tolerance checkpoint is defective in WAS patients. Frequency of polyreactive antibodies among mature naïve B cells from WAS patients. Antibodies from mature naïve B cells isolated from 4 WAS patients were tested by ELISA for reactivity against dsDNA, insulin and LPS. Polyreactive antibodies reacted against all 3 antigens. Dotted lines show ED38-positive control. Horizontal lines show cutoff OD405 for positive reactivity. For each individual, the frequency of reactive and non-reactive clones is summarized in pie charts, with the number of antibodies tested indicated in the center.



Supplemental Figure 6

Altered Treg phenotype in WAS patients. Expression levels of CD25, CD127, CD45RO and CTLA4 on CD4⁺FOXP3⁺Helios⁺ Treg and CD4⁺FOXP3⁺Helios⁻ non-Treg from HD controls and WAS patients are shown as mean fluorescence intensities (MFI) (CD25, CD127 and CTLA4) and percentage (CD45RO). Representative flow cytometry histograms of one HD control and one WAS patient are shown on the right. Differences were analyzed for statistical significance using Mann-Whitney test.



Supplemental Figure 7

Mature naive B cells from WAS patients post-GT express normal frequencies of polyreactive antibodies. Frequency of polyreactive antibodies among mature naive B cells from WAS patients after gene therapy. Antibodies from mature naive B cells isolated from 4 WAS GT-treated patients after gene therapy were tested by ELISA for reactivity against dsDNA, insulin and LPS. Polyreactive antibodies reacted against all 3 antigens. Dotted lines show ED38-positive control. Horizontal lines show cutoff OD405 for positive reactivity. For each individual, the frequency of reactive and non-reactive clones is summarized in pie charts, with the number of antibodies tested indicated in the center.

Table S1. Description of WAS patients treated by LV-mediated HSC-GT.

	WAS1* [¶]	WAS2* [¶]	WAS3* [¶]	WAS4 [¶]
WAS Mutation	Exon 10 c.961C>T (R321X)	Exon 10 c.1337_1338+9del	Exon 1 c.37C>T (R13X)	Exon 1 c.91G>A (E31K)
Pre HSC-GT, Zhu score	3	4	3	5
Pre HSC-GT, yrs at analysis	5.25	1.51	1.04	2.23
Pre HSC-GT, WASp+ lymphocytes (%)	4.3	1.5	4.5	3.0
Pre HSC-GT, WASp+ B cells (%)	3.9	0.8	17.5	2.9
Post HSC-GT, yrs of FU	2	1.5	1	1
Post HSC-GT, WASp+ lymphocytes (%)	54.9	69.1	66.7	64.9
Post HSC-GT, WASp+ B cells (%)	47	50.5	50.6	55.8

HSC-GT, Hematopoietic Stem Cell-Gene Therapy; Zhu Score, scoring system used to measure the severity of disease (1); Yrs, years; FU, follow up; c. coding DNA.

*, Patients already described in Aiuti et al., Science 2013 (30).

[¶], Patients and data already described in Castiello et al. (31).

Table S2: Repertoire and reactivity of antibodies from new emigrant B cells of HD30

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
neHD30 03	4-39	5-12	2	4	PSLSGYDYSLDY	12	3-20	3	QQYGSSPLFT	10	-	-	-
neHD30 04	3-15	/	/	3	GGGVGAFDI	9	2-28	5	MQALQTPT	8	-	-	C
neHD30 05	3-30-3	1-26	2	6	QSSPTGGGMDV	11	1-5	2	QQYNSYSYT	9	-	+	-
neHD30 10	4-4	3-9	3	5	VSNHILTNRLFDP	14	3-20	1	QQYGSSPRT	9	-	+	-
neHD30 12	1-2	2-15	2	3	HPIGYCSGGSCYGGAFDI	18	1-33	4	QQYDNLPT	8	-	-	-
neHD30 14	4-34	3-10	3	2	KGTMVRGVITPHWYFDL	18	2-28	1	MQALQTPQT	9	-	+	-
neHD30 16	3-30	/	/	6	EGVSHYYYGMDV	12	2-28	4	MQALQTPPT	9	-	-	-
neHD30 20	3-30	4-17	2	6	DIHRDYGDIETPNYYYYGMDV	21	1-9	1	QLNSYPLA	9	-	+	-
neHD30 24	3-72	/	/	6	DNRGMDV	7	4-1	4	QQYYSTPLT	9	-	-	-
neHD30 26#	1-18	3-16	2	4	NYDYIWGSYFTRGGY	15	1-33	3	QQYDNLPLFT	10			
neHD30 43	3-9	3-22	2	2	VDSSGLFGWYFDL	13	3-20	1	QQYGSSPQT	9	-	-	-
neHD30 45#	3-15	2-15	3	4	DPLIVVVVAATRDFDY	16	1-39	2	QSYSTPYT	9			
neHD30 02	4-34	3-10	2	4	GQDYGGSGSTADY	13							
neHD30 29	3-23	6-13	2	4	DEVSSSWLFGY	11							
neHD30 38	3-72	3-10	1	6	AGSWWFGETRYGMDV	15							
neHD30 41	3-23	5-12	2	4	VRGRPDSGYRFDY	13							
neHD30 44	1-69	2-15	2	6	GSYCSCGGSCYSSFYGGMDV	21							
neHD30 46	3-21	/	/	6	DDNLPIDYGGMDV	12							
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
neHD30 07	4-59	/	/	4	DRNFDY	6	3-1	2	QAWDSSTAV	9	-	-	-
neHD30 11	3-33	/	/	4	GGGGGDY	7	2-14	2	SSFTSITYVV	10	-	-	-
neHD30 19	3-11	1-7	2	4	DRGNNGRPNYNWYFFDY	18	2-14	1	SSYTSSSTLLYV	12	-	+	-
neHD30 21#	3-53	4-17	2	4	VGRYGDYAYTISRYYFDY	20	2-14	2	SSYTSSSTLGV	11			
neHD30 28	3-15	1-26	1	6	AGITKFWELPGYYYYGMDV	19	1-44	3	AAWDDSLNVVV	11	-	-	-
neHD30 30	3-23	2-21	3	6	GEGGVVTTQALMDV	15	1-44	2	AAWDDSLNGVV	11	-	-	-
neHD30 42	1-69	4-4	2	6	SDYSNYVLYDYGGMDV	19	2-8	2	SSYAGSNNLV	10	+	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S3: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 1 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS01 04 #	4-39	2-21	2	3	YLAYCGGDCYQPPPGSTWASAFDI	24	3-20	5	QQYGSSPLIT	10			
neWAS01 09	5-51	6-13	3	3	RIAAAGNAFDI	11	3-15	4	QQSNT	5	-	-	-
neWAS01 15	3-33	3-10	2	4	GSHGSGSYWAPYFDY	15	3-11	3	QQRSNWPLFT	11	-	-	-
neWAS01 17	3-66	2-15	2	6	DFQYCSCGGSCYSYGMDI	17	1-12	1	QQANSFPWT	10	-	-	-
neWAS01 25	3-7	2-2	3	6	DQVVPAAFSMEGWYGMVDV	20	1-39	1	QQSYSTRT	8	-	-	-
neWAS01 37	4-39	3-10	2	4	GANYYSGSYYPFDY	15	3-20	1	QQYGSSPGT	9	-	-	-
neWAS01 109	1-2	/	/	6	ELVGEGAYGMDV	12	1-39	3	QQSYSTPFT	9	-	-	-
neWAS01 113	1-69	1-26	2	4	LPGRYSGSFGYLDY	15	3-15	4	QQYNNWLT	8	+	+	-
neWAS01 138 #	1-69	/	/	4	SGTAETHFDY	11	3-15	1	QQYNNWPKT	9			
neWAS01 139	3-33	3-10	3	6	DYGEVRGVSVTYYGMDV	18	1-9	4	QQLNSYPLT	9	-	-	-
neWAS01 141	1-18	2-21	2	5	AVAYCGGDCYPTDTNWFDV	19	1-39	3	QQSYSTPFT	9	-	-	-
neWAS01 07							3-11	4	QQRSNWPLT	9			
neWAS01 08							1-5	1	QQYNSYPLT	9			
neWAS01 22							3-20	1	QQYGSSPWT	9			
neWAS01 27							3-20	5	QQYGSSPITFD	12			
neWAS01 29							2-28	1	MQALQTPPWT	10			
neWAS01 43							1-39	1	QQSYSTPRT	9			
neWAS01 48							3-20	2	QQYGSSPPYT	10			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS01 01	1-46	6-13	3	6	CMTNSIAAAGSWDYYYYYGMDV	22	3-1	3	QAWDSSTV	8	-	-	-
neWAS01 02	3-23	3-16	2	4	DSTLPSITYDYV	12	1-40	1	QSYDSSLGIV	11	-	+	-
neWAS01 03	3-15	3-22	2	4	GDRGTYYDSSGYRDY	16	9-49	3	GADHGSGSNFVRV	13	-	+	-
neWAS01 05	3-23	5-12	2	4	GSGGYDTYYFDY	12	2-11	1	CSYAGSSHV	9	-	-	-
neWAS01 14	4-61	2-15	2	2	ARWYCSCGGSCQDWYFDL	17	2-14	2	SSYTSSSTVV	10	-	-	-
neWAS01 32	4-39	3-22	2	5	DGNYYDSSGSGWVDFP	16	3-21	2	QVWDSSSDHVV	11	-	-	-
neWAS01 102	3-7	2-2	3	6	DEGDIVVPARYGMDV	16	3-21	2	QVWDSSSDHLVV	12	-	-	-
neWAS01 119	1-8	3-16	2	6	SGVDYVWGSYRPPHYYYYYGMDV	23	2-11	2	CSYAGSPVV	9	-	-	-
neWAS01 146	1-18	3-22	2	7	SLYYYDSSGSDY	12	3-1	3	QAWDSSTAR	9	-	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S4: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 2 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS02 01	3-49	1-26	2	3	DEGGSYYRGDAFDI	14	1-5	3	QQYNSYSFIT	10	-	-	-
neWAS02 14	4-4	3-16	2	5	DRGDDYVWGSYRTKPKWNWFDP	22	1-5	1	QQYNSYSGT	9	+	-	-
neWAS02 15	3-15	6-13	2	4	DLPSLGSSSWYGTEWGGDYF	20	4-1	2	QQYYSTPPYT	10	-	-	-
neWAS02 17	3-49	6-13	3	4	DLIAAAVKFDY	11	2-29	2	MQSIQLPLYT	10	-	+	-
neWAS02 20	3-15	2-2	2	6	VGVCSSSTSCYRGMDV	16	2-28	3	MQALQTLTLLFT	10	-	-	-
neWAS02 21	4-34	6-13	1	4	GPGQQLAWDY	10	1-39	3	QQSYSTPRT	9	-	-	-
neWAS02 25 #	3-23	5-5	1	4	TIQPSLPQISYFFDY	15	3-15	5	QQYNNWPFITFD	12			
neWAS02 28	4-59	1-26	2	4	TSGSYPYFFDY	11	3-11	4	QQRSNWLT	8	-	+	-
neWAS02 30	4-34	6-19	2	4	VNVGWGGPWSD	11	1-39	1	QQSYSTPRT	9	-	+	C
neWAS02 31	1-24	4-23	2	3	GNGGNSRQRKDAFDI	15	1-27	1	QKYNAPRT	9	-	-	-
neWAS02 11							1-33	3	QQYDNLPT	9			
neWAS02 16							1-39	1	QQSYSTPPT	9			
neWAS02 22							3-20	1	QQYGSSTWT	9			
neWAS02 26							1-39	4	QQSYSTPLT	9			
neWAS02 29							2-29	5	MQSIQLPITFD	11			
neWAS02 42							4-1	1	QQYYSTPPWT	10			
neWAS02 43							1-5	2	QQYNSYPYT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS02 02	3-9	1-26	3	6	GVGANPYYGMDV	12	2-33	3	CSYAGSSTWV	10	-	-	-
neWAS02 03	3-23	3-22	2	3	DLTEDYYDSSGSHDAFDI	18	1-44	2	AAWDDSLNGPV	11	-	-	-
neWAS02 04	3-30	2-21	2	4	DGLTLTDCGGDCYPHY	16	2-8	2	SSYAGSNNLHVV	12	-	-	-
neWAS02 07	5-51	5-12	2	4	TRGKSPSKYSGYDIDY	16	7-46	1	LLSYSGAYV	9	-	+	-
neWAS02 18	3-48	/	/	4	GGRSSDY	7	2-14	2	SSYTSSSLV	9	-	-	-
neWAS02 19	3-15	3-22	2	3	EEYYYDSSGYSAFDI	16	1-47	2	AAWDDSLSGRV	11	-	-	-
neWAS02 38	3-23	1-26	3	4	GGGVGATTPNFDY	14	2-14	2	SSYTSSSTLGV	11	-	-	-
neWAS02 39	1-2	2-15	3	3	DREVVVVAATRGIAFDI	18	2-14	2	SSYTSSSTHVV	11	-	-	-
neWAS02 40	3-48	/	/	4	DLLSEIDY	8	2-14	2	SSYTSSSTPV	10	-	-	-
neWAS02 45	3-20	6-13	3	3	VFHAPAAAAGSTDAFDI	17	1-44	1	AAWDDSLNGSYV	12	-	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S5: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 3 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS03 06	3-23	6-19	3	2	DLGSVAGTRRRYYWYFDL	17	3-20	5	QQYGSSPIT	9	-	+	-
neWAS03 07 #	3-9	3-10	2	6	VLYGSGKNFYYYGMDV	17	3-20	3	QQYGSSPIT	9			
neWAS03 10 #	4-39	3-22	2	3	ARYYYDSSGSADFID	14	1-5	5	QQYNSYSIT	9			
neWAS03 11 #	1-18	/	/	4	ERGTTGVVDY	9	3-11	2	QQREYT	6			
neWAS03 14	3-48	4-17	3	3	YTVTTRHAFDI	11	1-39	3	QQSYSTLFT	9	-	-	-
neWAS03 15	3-30	6-13	2	6	GTLSSILLWGMDV	13	2-28	2	MQALQTPYT	9	-	-	-
neWAS03 16	4-b	3-10	1	4	IWFGELLRPLGDY	13	3-20	1	QQYGSRT	8	+	-	-
neWAS03 17 #	1-8	1-7	2	4	ANY	3	1-16	5	QQYNSYPIT	9			
neWAS03 21	3-33	5-12	3	4	GGVVATITQLLFDY	14	1-16	3	QQYNSYPFT	9	-	+	-
neWAS03 23	4-39	6-19	3	3	AIQNSRIAVAGTGAFDI	17	3-20	1	QQYGSPPWT	9	-	-	-
neWAS03 24	4-39	3-10	2	4	LHGSVVDY	9	3-20	4	QQYGSPLT	8	-	-	-
neWAS03 26	3-11	3-22	2	4	DLYDSSGYLGY	13	1-6	4	LQDYNPLT	9			
neWAS03 27	3-7	5-12	3	3	AGGGVATIGSDAFDI	15	1-5	1	QQYNSYPWT	9	-	-	-
neWAS03 28	4-31	3-22	2	3	ARSNYDSSGPDADFID	16	1-5	1	QQYNSYPWT	9	-	+	-
neWAS03 29	1-3	3-10	3	4	SMVRGVPPFDY	10	1-33	3	QQYDNLHIFT	10	-	-	-
neWAS03 34	3-30	5-5	2	6	VFVSRFRPRGYSYQPPNTYYYYYGMVDV	28	2-28	4	MQALQTPLT	9	-	-	-
neWAS03 36 #	3-23	6-13	3	2	PTLGIAAAGPLSEDYWYFDL	20	3-11	5	QQRN WPPIT	12			
neWAS03 37	3-30-3	4-17	2	4	GPYDYGDPGSAY	13	4-1	1	QQYYSTLRT	9	-	-	-
neWAS03 38	3-15	2-8	2	4	DLGYCTNGVCYRPSYFFDY	19	1-17	4	LQHNSYPLT	9	-	+	-
neWAS03 41 #	3-21	6-13	3	4	ATLHEAAGNLDYFDY	15	3-11	2	QQRN WPPST	10			
neWAS03 46 #	3-23	2-15	3	4	DMGSDIVVVAATPDY	16	1-8	1	QQYNSYPRT	10			
neWAS03 01							1-39	2	QQSYSTPYT	9			
neWAS03 18							3-15	1	QQYNNWPRT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS03 03	3-7	3-10	2	4	DQYYYGSGSYLDY	14	2-11	1	CSYAGSYTRYV	11	-	-	-
neWAS03 13	3-15	1-26	2	4	DPNSGSFPPGDY	12	2-14	3	SSYTSSSPWV	10	-	-	-
neWAS03 22	3-30	5-5	3	6	DQSSGTAMAFYGMVDV	15	1-51	3	GTWDSLSAGV	11	-	-	-
neWAS03 25	4-39	3-10	3	5	DSPITMAHWFDP	12	2-8	2	SSYAGSNKPVV	11	-	-	-
neWAS03 30	3-7	4-4	2	6	SLYSNYVSLSYGMVDV	15	2-14	2	SSYTSSSTLV	10	-	-	-
neWAS03 35	3-30	6-19	3	4	DQAVAGHLFDY	11	2-14	1	SSYTSSSYV	9	-	+	-
neWAS03 39	3-30	/	/	3	DTANFVTYEKIKGAFDI	17	2-11	1	CSYAGSYTYV	10	-	-	-
neWAS03 43	5-51	5-5	2	3	HFPSPPEGYSYARTLDAFDI	20	2-14	1	SSYTSSSTLYV	11	-	-	-
neWAS03 45	3-53	2-15	2	3	GRPYCSGGSCVDAFDI	16	3-21	2	QVWDSSTDHVV	11	-	+	-
neWAS03 08							1-51	3	G TWDSLSAGV	12			
neWAS03 19							1-51	2	CTWDSLSAVV	11			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S6: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 4 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	V _κ	J _κ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS04 14	3-30	2-15	2	4	DLGYCSGGSCYRGDDY	16	3-15	2	QQYNNWPRYT	10	-	-	-
neWAS04 15	3-15	1-26	2	6	SGSYTKYYYYGMDV	14	1-39	4	QQSYSTPLT	9	-	-	-
neWAS04 18 #	4-34	3-10	1	5	GRREFGELLSLNWFDP	16	3-15	5	QQYNNWPPITFD	12			
neWAS04 25	4-30-4	/	/	4	ETRAKHHLEGGNDY	14	1-39	1	QQSYSTPRT	9	-	-	-
neWAS04 38	3-21	3-22	2	4	VHSSGYPDY	9	3-20	4	QQYGSSPPT	9	-	-	-
neWAS04 44	3-49	2-2	2	6	GRGGGTTCPYGMDV	14	2-28	3	MQALQTPQT	9	-	+	-
neWAS04 113	4-39	3-22	2	4	GIAERYDSSGYYYYFDY	18	3-20	3	QQYGSSPLFT	10	-	-	-
neWAS04 114	3-23	6-6	2	4	PYSSSSVPSY	12	3-11	4	QQRSNWPLT	10	-	-	-
neWAS04 124	3-9	4-17	2	4	SHGDYEPFDY	10	3-15	1	QQYNNWPGT	10	-	-	-
neWAS04 125	4-39	3-10	2	4	DSGYGSGSYVNDY	14	3-20	3	QQYGSSPGFT	10	-	-	-
neWAS04 126	3-23	/	/	4	DRGYSISHFDY	11	3-11	4	QQRSNWPT	8	-	-	-
neWAS04 130	4-39	6-19	3	4	EGEAAVAGSFDY	12	4-1	2	QQYYSTPPT	9	+	+	-
neWAS04 137 #	1-18	6-19	2	6	RPLPDLEPYSSVATDYYYYGMDV	24	2-28	3	MQALQTPT	8			
neWAS04 142 #	3-30	6-13	3	4	VPGIAAAGTLNYFDY	15	3-11	2	QQRNNWLYT	9			
neWAS04 143	3-21	3-22	2	4	DRYDSSGLIDY	11	1-5	2	QQYNSYSGT	9	-	+	-
neWAS04 144	3-66	3-3	2	4	DGGSREYDFWSGYDY	16	1-5	2	QQYNSYSYT	9	-	+	-
neWAS04 106	4-b	6-13	3	4	ASSIAAAGYFDY	12							
neWAS04 127	4-59	3-3	3	6	DQTIFEDGVGYYYGMDV	18							
neWAS04 128	4-34	3-10	2	3	EWNYGSGKRTNDAFDI	16							
neWAS04 132	3-21	4-17	3	2	DAVTVTTPLWYFDL	15							
neWAS04 133	3-23	2-21	3	4	SQRGVVVTASDY	12							
neWAS04 141	1-24	1-26	2	4	GARD	4							
	VH	D	RF	JH	CDR3(aa)	Length	V _λ	J _λ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS04 10	3-30	2-2	2	4	EGLGYCSSTSCYFFDY	16	1-51	2	GTWSSSLSGVV	11	-	-	-
neWAS04 13	1-18	3-10	1	4	DKNRGWFGGSPVDY	14	2-18	2	SLYTSSSTV	9	-	+	-
neWAS04 28	4-39	6-13	1	4	QQQLAPDY	8	1-44	3	AAWDDSLNGFWV	12	-	-	-
neWAS04 31	3-43	2-15	3	6	DGGAVVAARYYYYYGMDV	19	2-14	2	SSYTSSSTV	10	-	-	-
neWAS04 102	3-30	/	/	6	DSVAEWGYHYYYGMDV	17	1-51	3	GTWSSLSAGV	11	-	-	-
neWAS04 104	3-15	4-23	2	4	GDGRGYGGNPFDY	13	2-14	1	SSYTSSSTV	9	-	-	-
neWAS04 109	3-15	5-12	3	4	DLIIVATISSDY	12	1-47	2	AAWDDSLSGRV	11	-	-	-
neWAS04 115	1-18	3-9	1	6	ERYFDYYYYYYGMDV	16	1-44	1	AAWDDSLNGYV	11	-	-	C
neWAS04 116	4-34	3-10	3	3	SARRVRGDRADAFDI	15	1-51	2	GTWSSSLRGVV	11	-	+	-
neWAS04 117	3-30	3-3	3	4	DRQFGVVIITSVGNFDY	18	3-21	7	QVWDSSSDQHAV	12	-	-	-
neWAS04 134 #	1-2	4-17	2	3	DGDYGDYGTAFDI	14	1-47	3	AAWDDSLSGRV	11			
neWAS04 135	4-34	/	/	4	GLHLVGAEDY	10	2-11	1	CSYAGSYKV	9	-	-	-
neWAS04 140	1-24	/	/	4	DLGRRRTWKVDY	12	2-18	3	SLYTSSSI	8	-	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S7: Repertoire and reactivity of antibodies from mature naïve B cells of HD30

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnHD30 56	3-33	6-19	3	4	DLQGIAVAGTTATGFDY	17	2-28	2	MQALQTPPT	9	-	-	-
mnHD30 58	3-30-3	3-3	2	5	DGVQEYYDFWSGYYGNWFDP	20	3-20	1	QQYGSSPPRT	10	-	-	-
mnHD30 66	1-69	/	/	3	DDQDYLRAFDI	11	1-5	1	QQYNSYST	8	-	-	-
mnHD30 67	1-69	5-24	3	4	VGGLEMATPLDY	13	3-11	4	QQRSNWPQLT	10	+	-	-
mnHD30 70 #	5-51	4-4	2	5	LLAGNDYSNPFDP	13	3-11	5	QQRSNWPPLIT	11			
mnHD30 71	3-13	2-15	2	4	GWGYCSGGSCYGFYD	15	1-9	4	QQLNSYPALT	10	-	-	-
mnHD30 72	3-13	3-10	2	6	AWKGSCKYYYGMDV	14	2-40	2	MQRIEFPYT	9	-	-	-
mnHD30 75	4-59	3-10	1	4	GSPHTIWFGESPPPLLDY	18	1-8	1	QQYYSYPQT	9	+	+	C
mnHD30 78	4-31	2-21	3	5	SGGRHIVVTAIPPFDP	17	1-5	1	QQYNSYST	8	-	-	-
mnHD30 83	4-31	/	/	5	VPGKGGWFDP	10	1-27	4	QKYNSAPLT	9	-	-	-
mnHD30 93	3-11	3-22	2	4	VGIYDSSGYLYFDY	15	1-6	3	LQDYNYPFT	9	-	-	-
mnHD30 95	1-69	3-10	2	4	GGSGSYNVLY	11	1-33	5	QQYDNLAF	8	-	-	-
mnHD30 49	3-49	/	/	4	DWPWEPNYFDY	12							
mnHD30 52	3-30-3	2-21	2	2	WELAYCGGDCHTAGAFDI	18							
mnHD30 57	1-18	/	/	4	DVGWKFDY	8							
mnHD30 88	3-30	2-2	3	4	EQVPAAMFDY	10							
mnHD30 92	3-30	3-10	1	4	DSLWFGELDY	10							
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnHD30 51	3-21	3-3	1	5	GLEWLLPPTFDP	12	2-14	1	SSYTSSTLYV	11	-	-	-
mnHD30 59	4-31	/	/	4	HYSYGAWDFNY	10	3-1	2	QAVDSSTVV	9	-	-	-
mnHD30 60	3-53	3-9	2	3	TTTISKTLDAFDI	13	3-21	3	QVWDSSTDHPV	11	-	-	-
mnHD30 68	3-30	5-12	2	4	DLSHSGYDLGIDY	13	2-14	1	SSYTSSTLV	10	-	-	-
mnHD30 76	7-4-1	1-26	2	4	RSGSYLQDFDY	11	3-21	3	QVWDSSTDHPV	11	-	+	-
mnHD30 80	3-15	1-26	3	5	DPVFIVGRASNWFDP	15	3-25	3	QSADSSGSPWV	11	-	+	-
mnHD30 85 #	1-2	/	/	6	ERIRPHLGRSGMDV	14	2-14	1	SSYTSSTVHYV	12			
mnHD30 87	1-69	3-10	1	3	MPEGELLSAHDAFDI	15	2-11	3	CSYAGSYHWV	10	-	-	-
mnHD30 91	3-74	6-19	2	3	SSGWTGAFDI	10	3-1	2	QAWDSSTVV	9	-	+	N+C

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S9: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 2 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS02 49	4-34	3-16	2	6	GPHTAGRFYVWGSYHDSYGMDV	21	1-5	4	QQYNSYPLT	9	+	+	C
mnWAS02 52	4-4	4-23	1	2	DRSELRWNNWYFDL	14	1-13	4	QQFNSYPLT	9	+	+	-
mnWAS02 76	3-30	6-19	3	3	DLEDWLAVAGSGAFDI	16	1-39	2	QQSYSTQYT	9	-	+	-
mnWAS02 77	3-30	6-19	3	6	DPSAVAGEGYYYYYGMDV	18	1-33	4	QQYDNLPLT	9	-	-	-
mnWAS02 85	4-59	6-13	2	3	DQDGYSSWSRAFDI	15	3-20	4	QQYGSSPLT	9	-	+	-
mnWAS02 87	3-53	6-19	3	3	ARRGAVAGFQDAFDI	15	1-5	3	QQYNSPLFT	9	-	+	-
mnWAS02 90	3-74	/	/	4	LLQYDY	6	1-39	1	QQSYSTPRT	9	-	+	-
mnWAS02 70							3-20	1	QQYGSSQWT	9			
mnWAS02 74							2-28	1	MQALQTSWT	9			
mnWAS02 92							1-5	2	QQYNSYPYT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS02 51	3-49	3-16	1	4	VVGEKLESLYYFDY	14	1-44	3	AAWDDSLNGWV	11	-	-	-
mnWAS02 53	3-13	4-4	2	4	GLGGYSNYVDY	11	7-43	3	LLYYGGARV	9	-	-	-
mnWAS02 55	4-59	/	/	4	VGRGPMADFDY	11	2-14	2	SSYTSSSTRLV	11	-	-	-
mnWAS02 57	1-18	7-27	3	4	DLTGDDYDY	8	1-40	1	QSYDSSLGSGV	12	-	-	-
mnWAS02 63	5-51	3-22	2	5	TYYYDSSGYTTGRWFDP	17	1-51	1	GTWDSSLAYV	11	-	-	-
mnWAS02 65	4-34	5-5	2	3	RGYSYGHAFDI	11	2-14	3	SSYTSSSTLWV	11	-	-	-
mnWAS02 78	4-34	3-10	3	3	GITMVRGVIPGYAFDI	16	1-51	2	GTWDSSLAVV	11	-	-	-
mnWAS02 82	5-51	/	/	4	APQRGVPEFDY	11	2-11	1	CSYAGSYTYV	10	-	+	-
mnWAS02 84	4-34	4-23	2	3	SDYAAFDI	8	2-14	2	SSYTSSSTGV	10	-	-	-
mnWAS02 86	3-48	2-2	3	6	DRVVPAARNYYYYGMDV	17	3-25	3	QSADSSGTYRV	11	+	+	C
mnWAS02 89	3-30	3-16	2	4	DGLGVWGSQTSYD	13	2-14	3	SSYTSSSTLV	10	-	-	-
mnWAS02 94	1-69	4-17	2	6	ESLYGDQETRYYYGMDV	17	1-51	3	GTWDSSLAWV	11	-	-	-
mnWAS02 95 #	1-18	3-10	1	3	VSVLLWFGGSRPDTSGAFDI	20	2-14	3	SSYTSSSTGV	10			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S10: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 3 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS03 49	1-18	/	/	3	ERVTVTSEGAFDI	13	4-1	2	QQYYSTPYT	9	-	-	-
mnWAS03 53 #	4-34	3-10	1	4	RGFGEYPPFDY	10	3-20	1	QQYGSSPWT	9			
mnWAS03 54	3-15	6-13	3	6	DRLSIAAAGTPNGPYYYGMDV	22	1-33	2	QQYDNLPLYT	9	-	-	-
mnWAS03 56	3-33	/	/	4	DLEGLSRFGETGYSSSWALGY	21	1-39	4	QQSYSTPPLT	10	-	+	C
mnWAS03 65	3-20	/	/	6	DRAPYYYYGMDV	12	1-39	3	QQSYSTPQEFT	11	+	+	-
mnWAS03 67	1-3	6-13	2	5	DANSSSWPDHNPSPNNWFDP	21	1-5	1	QQYNSYPT	8	-	-	-
mnWAS03 69	3-23	3-10	3	4	DRQTFMVRGVIDGY	14	4-1	1	QQYYSTPRT	9	+	+	-
mnWAS03 70	4-59	2-21	3	3	EFDGPSVVVTGHDAFDI	17	3-20	4	QQYGSSPPLT	10	-	+	-
mnWAS03 77 #	4-34	6-13	1	2	GFQQQLAQGYFDL	13	2-29	1	MQSIQRPWT	9			
mnWAS03 85	3-48	5-5	2	6	DMSGYSYGYYYGMDV	17	3-20	2	QQYGSSSYT	9	+	+	N
mnWAS03 86	5-51	/	/	4	NAFTPAPGVG	10	1-17	2	LQHNSYPYT	9	-	+	-
mnWAS03 88	3-9	/	/	4	SHQSGVIF	8	3-11	4	QQRSNWLT	8	-	-	-
mnWAS03 93 #	4-31	3-22	2	4	GRVYYYYDSSGSAFDY	16	1-5	1	QQYNSYPWT	9			
mnWAS03 28							1-8	4	QQYYS YPLT	10			
mnWAS03 29							1-27	4	QKYNSAPLT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS03 51	3-9	6-19	3	6	DIRAVAGTNYYYGMDV	17	1-51	3	GTWDSLSAGV	11	-	-	-
mnWAS03 57	5-51	3-10	2	3	GRGDYGSYSFPAGAFDI	18	1-44	2	AAWDDSLNGLVV	12	-	-	-
mnWAS03 62	3-23	3-10	2	4	DSERTHSLDYGSGSYDPFDY	22	3-21	2	QWWDSSSDHRRV	12	-	+	-
mnWAS03 63	4-39	3-10	2	4	EHGSGYSLDY	11	2-14	3	SSYTSSSTLWV	11	-	-	-
mnWAS03 68 #	1-69	3-10	2	4	EVEYYYGSGSYTRGRFDY	19	3-1	2	QAWDSSTVV	9			
mnWAS03 74	3-33	3-10	3	4	DGDGITMVRGVTPRYFDY	18	2-14	1	SSYTSSNSYV	10	-	+	-
mnWAS03 75	3-23	3-22	2	3	DLYDSSGYWGDDAFDI	16	2-14	3	SSYTSSSTRV	10	-	-	-
mnWAS03 78 #	3-9	1-26	2	4	DMLSGSRPAYFDY	13	2-14	1	SSYTSSSTPLYV	12			
mnWAS03 80	3-30	1-26	3	4	LVGATQGDY	9	2-14	1	SSYTSSSTL	9	-	-	-
mnWAS03 82	3-20	2-15	2	4	DLARYCSGGSCYPLGPEFDY	20	1-51	2	GTWDSLSAVV	11	-	-	-
mnWAS03 91	3-9	1-26	3	6	DFNRGRGSGGMDV	13	3-1	2	QAWDSSTAV	9	-	-	F

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S11: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 4 before GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS04 53	4-34	6-6	2	4	GFSSSPNDY	9	3-20	4	QQYGSSPLT	9	-	+	-
mnWAS04 64	3-49	3-3	1	2	DEVVLRFLFWLPTGSDL	17	1-33	2	QQYDNLPLT	9	+	+	-
mnWAS04 73	4-39	6-19	3	6	GGIAVAGMIINYYGMDV	17	1-5	1	QQYNSYPWT	9	-	+	C
mnWAS04 74	3-7	5-5	3	6	AWGAAMVTRDYGMDV	15	1-16	1	LQYKTYPRT	9	-	+	-
mnWAS04 90	3-23	6-6	3	5	PPVRIAARVR	10	1-33	2	QQYDNLPPYT	10	+	+	-
mnWAS04 94	3-11	6-19	3	4	DQVAPLLRAKSGAIDY	16	1-39	1	QQSYSTPPWT	10	-	+	-
mnWAS04 162 #	4-34	2-15	3	4	AAAVSVVDETPYYFDY	17	3-20	5	QQYGSSPITFD	11			
mnWAS04 164	1-2	4-17	2	3	DGDYGDYGTAFDI	14	3-20	4	QQYGSSL	7	-	-	-
mnWAS04 165	3-30	4-17	2	3	DPFDYGDYPDAFDI	14	1-5	2	QQYNSYPYT	9	-	-	-
mnWAS04 177	3-23	2-21	3	4	DSSGDIVVTTDDY	13	3-11	2	QQRSNWPPVT	10	-	-	-
mnWAS04 87	3-15	3-10	1	5	DLSSVVGKWWFGDV	14							
mnWAS04 91	3-30	/	/	4	GGHATLWGGFDY	12							
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS04 49	3-30	6-19	3	4	EGEAAVAGSFDY	12	2-23	2	CSYAGSSTLVV	11	-	-	-
mnWAS04 51	4-34	3-3	2	3	PSGNYDFWSGYPIFAFDI	18	3-21	3	QVWDSSTLHWV	11	+	+	C
mnWAS04 57	3-48	3-10	1	3	QDRVPWFGEYGVFDI	16	1-44	1	AAWDDSLNGLF	11	-	-	-
mnWAS04 80	4-59	3-3	1	4	SLKKWLLSY	9	1-51	2	GTWDSLSLGVV	11	-	-	-
mnWAS04 186	3-66	2-2	3	4	GSDIVVPAALTFDY	15	3-1	3	QAWDSSTLVL	10	-	-	-
mnWAS04 188	1-69	/	/	6	GNYYYYGMDV	10	2-14	2	SSYTSSTLVV	11	-	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S12: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 1 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	V _κ	J _κ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS01p 01	3-23	3-22	2	1	SDSSGYYSIYFQH	13	1-12	4	QQANSFPLT	9	-	-	-
neWAS01p 06 #	1-69	3-22	3	3	GAGDITMIVQEEGAFDI	17	3-20	4	QQYGSSPLT	9			
neWAS01p 08	3-53	4-23	2	4	DYGGKRGDY	9	3-11	2	QQRSNWPPHT	10	-	-	-
neWAS01p 11 #	3-23	2-2	2	4	DRGCSSTCTYYFDY	15	1-33	2	QQYDNLPLT	9			
neWAS01p 12	1-46	5-5	2	4	DRVEYSYGPFDY	12	3-20	2	QQYGSSPPLMYT	12	-	-	-
neWAS01p 13	4-30-2	4-17	2	4	VGRSSDYGDYRGPSHYFDY	19	1-39	4	QSYSTLLT	9	-	+	-
neWAS01p 14 #	4-61	/	/	5	HRPVAGINWFDP	12	1-39	2	QSYSTPYT	9			
neWAS01p 15	3-30	6-19	2	6	VGSSGWYAPPYYGMDV	16	1-39	4	QSYSTPLT	9	-	+	-
neWAS01p 19	3-15	1-26	2	6	ETHNSGSLYYYYYGMV	19	1-39	4	QSYSTPLT	9	-	+	-
neWAS01p 25 #	1-46	5-5	2	4	DRVEYSYGPFDY	12	1-27	4	QKYNAPLT	9			
neWAS01p 36	7-4-1	/	/	3	DTWPNAFDI	9	4-1	1	QQYYSTPRT	9	-	-	-
neWAS01p 38	3-15	6-25	3	4	DRIGIAASRGPGRSNNFDY	20	3-11	4	QQRSNWPPLT	10	+	+	-
neWAS01p 40	3-15	2-2	3	6	DEVIVVPAATNYYYYGMV	20	3-20	3	QQYGSSPH	8	-	-	-
neWAS01p 28	4-39	3-16	1	4	GGGELSLPIDY	12							
neWAS01p 32	1-69	3-22	2	4	DRDYDSSGYVDGFDY	16							
neWAS01p 35	1-24	4-23	2	3	PKPRYGGNSVSSPAFDI	17							
neWAS01p 33							3-15	1	QQYNNWPMA	9			
neWAS01p 39							3-20	4	QQYGSSPPLT	11			
	VH	D	RF	JH	CDR3(aa)	Length	V _λ	J _λ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS01p 02 #	3-30	3-22	2	4	AFHGDSSGYSCDY	14	1-51	3	GTWDSLSAGV	11			
neWAS01p 17 #	7-4-1	6-19	3	4	DHHAVAGTALTPGLGH	16	1-44	3	AAWDDSLNGWV	11			
neWAS01p 20	3-33	/	/	6	GGAGYHYYYGMV	13	3-21	3	QVWDSSSDHWV	11	-	-	-
neWAS01p 24	3-30	/	/	4	SFDY	4	2-14	3	SSYTSSTDWV	11	+	-	-
neWAS01p 26 #	3-48	3-16	3	2	EYMITFGGVRDANWYFDL	18	2-23	2	CSYAGSSTFE	10	-	-	-
neWAS01p 30	4-30-4	5-5	2	3	GGYSYDAQGAFDI	13	2-14	2	SSYTSSTV	10	-	-	-
neWAS01p 31 #	3-30-3	3-10	3	4	VGSVTMVRGVDY	12	6-57	3	QSYDSSTWV	9			
neWAS01p 43	7-4-1	2-2	3	6	DRLFVVVPAAYGMV	15	2-23	2	CSYAGSSTLV	10	-	+	-
neWAS01p 45	1-18	4-17	3	3	RATVTTTGAFDI	12	2-23	3	CSYAGSSTWV	10	-	-	-
neWAS01p 46	3-30	/	/	6	LGYYGMV	8	2-23	1	CSSAGSTSYV	10	-	+	-
neWAS01p 47	1-18	6-13	2	4	LGHSSSWTFDY	11	2-14	3	SSYTSSTWV	10	-	-	-
neWAS01p 27							1-47	3	AAWDDSLSVNWV	12			
neWAS01p 37							2-14	3	SSYTSSSIWV	10			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S13: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 2 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS02p 02 #	1-2	4-4	2	4	VDRDSNSSLGFDY	13	1-39	1	QSYSTPRT	9			
neWAS02p 04	3-30	3-22	2	4	GGYDSSGSWDY	11	3-15	1	QYNNWPPWT	10	-	-	-
neWAS02p 05	3-11	3-10	2	6	NGSGSYSLYYYGMDV	17	1-9	1	QLNSYPWT	9	-	-	-
neWAS02p 09	4-59	6-19	2	3	GGSSGADAFDI	11	3-20	4	QFGKSLT	8	-	-	-
neWAS02p 12	3-15	/	/	4	GVEAYPLPDY	10	1-8	2	QYYSYPRT	10	-	-	-
neWAS02p 14	3-11	1-7	2	4	PSGPDNWNVY	9	3-15	3	QYNNWPLFT	11	-	+	-
neWAS02p 20	4-59	4-4	3	6	EVTTCGMDV	9	1-39	2	QSYSTPYT	9	-	-	-
neWAS02p 28 #	1-58	6-6	3	4	DGVIAARGLPDY	13	3-15	2	QYNNWPTY	9			
neWAS02p 30	3-21	1-7	2	4	DFGSIWNVPSDY	13	1-17	2	LQHNSYPYT	9	+	+	C
neWAS02p 32 #	3-49	6-13	3	4	GGPRAIAAGTKGEGVFDY	19	1-16	5	QYNSYPIT	9			
neWAS02p 33	3-53	/	/	2	DPREGIYWYFDL	12	3-20	2	QYGSPPYT	10	-	-	-
neWAS02p 34	3-74	5-5	3	4	VGVDTAMVTPVDY	13	3-15	2	QYNNWPLMYT	11	-	+	-
neWAS02p 37	3-7	3-10	2	2	EYSGSYPHWYFDL	15	3-20	1	QYGT	5	-	+	-
neWAS02p 39	3-33	4-23	2	3	EVSPRYGGNSGVSAFDI	17	3-20	2	QYGSPPMYT	10	-	-	-
neWAS02p 18	3-74	2-2	3	6	DETAEDIVVPAIYYGMDV	20							
neWAS02p 43	1-18	5-12	2	5	DNGGWDSGYDFGGWFDP	17							
neWAS02p 46	3-30	2-2	3	6	DIVVPAALYYYGMDV	17							
neWAS02p 47	4-59	2-2	2	3	GYCSSTSCRASNGAFDI	18							
neWAS02p 17							3-15	2	QYNNWPPDT	10			
neWAS02p 24							3-20	2	QYGSPPYT	9			
neWAS02p 26							3-20	1	QYGSLLPT	9			
neWAS02p 27							3-20	3	QYGSPPFT	10			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS02p 03	1-69	/	/	6	DHRQPQAYYYYGMDV	16	2-14	1	SSYTSSTRV	10	-	-	-
neWAS02p 06 #	1-46	5-5	2	4	GTFLPGYSYAPGYFDY	17	2-14	1	SSYTSSTYV	10			
neWAS02p 13	3-30	2-2	3	4	DIGVVAASCIDY	14	1-47	2	AAWDDSLGGRVV	13	-	-	-
neWAS02p 21	4-61	3-10	2	4	YGSGSYTRDTAVAGYLDY	18	1-44	1	AAWDDSLNGYV	11	-	-	-
neWAS02p 22	3-15	3-22	3	4	SRITMIVGY	9	1-44	2	AAWDDSLNGVV	11	-	+	-
neWAS02p 23	3-11	2-15	2	6	VGYCSCGGSCYPTGSYYYGMDV	21	2-14	2	SSYTSSTLV	10	-	-	-
neWAS02p 25	3-53	4-23	3	5	SDPTTVVPQFDP	12	2-23	3	CSYAGSSTFG	10	-	+	-
neWAS02p 35	3-7	3-22	2	3	EMHYDSELGVNAFDI	17	2-14	1	SSYTSSTLYV	11	-	-	-
neWAS02p 38	3-13	6-19	2	4	ANYSSGWYGVYD	12	2-23	2	CSYAGSSTLV	10	-	-	-
neWAS02p 44	4-4	5-5	2	4	SDGYSYGFYD	10	2-23	1	CSYAGSSTYV	10	-	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S14: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 3 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	V _κ	J _κ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS03p 03	4-39	3-22	2	4	DLLNYDSSGY	10	3-20	1	QQYGSSPPWT	10	-	-	-
neWAS03p 06	3-30-3	4-23	2	4	RQRVWVDYGAAVDY	14	3-20	5	QQYGSSPIT	9	-	+	-
neWAS03p 08	3-73	1-26	1	4	PGRDLELLIDY	11	1-39	1	QQSYSTPWT	9	-	-	-
neWAS03p 10	3-30	6-19	3	4	PTVGVGLAVAAPFDY	15	1-5	2	QQYNSYST	8	-	+	-
neWAS03p 14	3-30	/	/	3	ARRPGTSGAADAFFDI	15	1-39	1	QQSYSTPQT	9	-	+	-
neWAS03p 18	3-30	6-6	3	6	GGIAARSYYYGMDV	14	1-12	3	QQANSFPFT	9	-	+	-
neWAS03p 25	3-49	6-19	3	1	GAVAGLEYFQH	11	3-15	3	QQYNNWPYS	9	-	-	-
neWAS03p 30	1-18	6-13	3	4	KPIAAAGTGWVVDY	14	3-15	1	QQYSNWPQT	9	-	+	-
neWAS03p 35	4-39	6-19	2	4	FHPYSGWLADY	11	1-5	1	QQYNSYST	8	-	+	-
neWAS03p 38 #	3-30-3	2-2	3	2	AAIVVPAAMPAYWFDL	17	3-15	4	QQYNNWPPLT	10			
neWAS03p 42	3-15	5-24	3	4	GAQMATINQAYYFDY	15	3-15	2	QQYNNWPPYT	10	+	-	-
neWAS03p 45	1-58	2-2	3	4	VAHIVVPAAPGY	13	3-20	1	QQYGSSPWT	9	+	+	C
neWAS03p 07	4-31	3-22	2	4	SSGYDSSGSYSDY	13							
neWAS03p 17	3-48	4-17	2	4	NYGGDY	6							
neWAS03p 20	3-15	/	/	2	INTPPTIAHWYFHL	14							
neWAS03p 21	3-49	3-22	2	4	VGDLTYYYDSSGYIDFDY	18							
neWAS03p 28	4-b	2-15	3	3	HTVVAATPHAFDI	14							
neWAS03p 15							1-39	1	QQSYSTPRT	9			
neWAS03p 34							1-5	5	QQYNSYSIT	9			
	VH	D	RF	JH	CDR3(aa)	Length	V _λ	J _λ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS03p 01	3-21	3-22	2	3	AHIESSYDSSVNAFDI	16	1-40	2	QSYDSSLGQV	11	-	-	-
neWAS03p 02	3-23	2-15	2	4	DSVPFGGGSCYGH	13	1-40	2	QSYDSSLGQV	11	-	-	-
neWAS03p 04	5-51	2-15	2	4	LLGYCSGGSCYGFDFY	16	1-44	3	AAWDDSLNGPV	11	-	-	-
neWAS03p 05	3-33	3-10	2	4	DLIHPFYGSGSYNGLGY	18	2-5	3	CSYTSSATWV	10	-	+	-
neWAS03p 06	3-30-3	4-23	2	4	RQRVWVDYGAAVDY	14	1-44	2	AAWDDSLNGHV	12	-	+	M
neWAS03p 11	1-3	6-19	3	5	VAVAGTDYLQYNWFDP	16	1-40	3	QSYDSSLGQV	11	-	-	-
neWAS03p 13	7-4-1	2-15	2	4	NLGYCSGGSCRLAY	15	1-44	3	AAWDDSLNGWV	11	+	-	-
neWAS03p 15	3-9	3-10	3	3	DTRTITMVRSPFDI	15	2-14	2	SSYTSSSTLV	10	-	-	-
neWAS03p 17 #	3-48	4-17	2	4	NYGGDY	6	1-44	7	AAWDDSLNGAV	11			
neWAS03p 19	3-15	6-13	3	6	ARIAAAPYYYYYGMV	17	2-14	2	SSYTSSSTHV	11	-	-	-
neWAS03p 22 #	3-30	2-15	2	4	GGNEPPSYCSGGSCYSYGFDFY	21	2-11	1	CSYAGSYTFV	10			
neWAS03p 27 #	3-23	2-21	2	5	VGSHCYEV	8	2-11	1	CSYAGSYTFV	10			
neWAS03p 29	3-11	4-23	2	4	ADYGGNSAGLQNFDFY	15	2-14	1	SSYTSSSTLKV	11	-	-	-
neWAS03p 30	1-18	6-13	3	4	KPIAAAGTGWVVDY	14	1-51	3	GTWDDSLSAGE	11	-	-	-
neWAS03p 31	3-30	6-13	2	4	GLSGISSFNLDY	12	2-14	1	SSYTSSSTLV	10	-	-	-
neWAS03p 32	3-23	2-15	3	4	DLFTEPSDIVVVAATPLDY	20	1-51	2	GTWDDSLSAGV	11	-	-	-
neWAS03p 36	3-13	6-19	3	3	VRAGSADAFDI	11	1-44	7	AAWDDSLNGPV	11	-	-	-
neWAS03p 37	3-30	2-21	2	6	DILRTNCGGDCYSPYYGMDV	20	1-51	2	GTWDDSLSAGV	11	-	+	-
neWAS03p 39 #	3-30	3-10	2	6	CSYSGSYTANYYYYYGMDV	20	2-8	1	SSYAGSNNLS	10			
neWAS03p 41	4-59	1-26	2	4	GPGVSGSYSFDFY	12	2-14	3	SSYTSSSTWV	10	-	-	-
neWAS03p 48	4-59	4-17	2	3	EGWDYGDYHGAFDI	14	1-44	3	AAWDDSLNGWV	11	-	-	-
neWAS03p 12							3-21	1	QWDDSSSDHPGYV	13			
neWAS03p 44							7-43	2	LLYGGDRV	9			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S15: Repertoire and reactivity of antibodies from new emigrant B cells of WAS patient 4 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS04p 01 #	3-21	6-6	3	5	GGIWGIAARPSWFDP	15	1-39	5	QQSYSTPPTT	10			
neWAS04p 04	3-23	1-26	3	4	APYRSSWVGAVHPDY	16	2-30	4	MQGTHWPLT	9	-	-	-
neWAS04p 09 #	3-11	6-6	3	4	SSIAEDY	7	3-20	4	QQYGSSPRLT	10			
neWAS04p 14 #	3-9	3-3	2	5	GDPWSDSPGFDP	12	1-17	2	LQHNSYPYT	9			
neWAS04p 16 #	5-51	2-15	2	4	GACSGGSCQGLLDY	14	1-5	1	QQYNIYWTAQAQ	11			
neWAS04p 22 #	3-30-3	5-5	3	5	TLYTAMVTRLGP	12	3-15	1	QQYNNWPTWT	10			
neWAS04p 27	1-24	1-26	2	4	DLSSGSYGKGGGFDP	15	1-5	1	QQYNSYSWT	9	+	+	C
neWAS04p 28	4-b	5-5	3	4	DTAMVTRFY	10	3-11	5	QQRSNWPPIT	10	-	+	-
neWAS04p 29 #	3-7	6-19	3	3	EGVAVAGTGAVAFDI	15	3-15	4	QQYNNWPPPLT	10			
neWAS04p 30	3-21	1-26	2	1	VGNSGSYYNGYFQH	14	3-20	2	QQYGSSPRT	9	-	+	-
neWAS04p 32	3-15	3-22	2	4	ASLDYDSSGYQDFDY	15	1-8	4	QQYYSYPLT	9	-	+	-
neWAS04p 33	3-30-3	3-3	2	4	DFWSGYYFDY	10	1-27	3	QKYNAPFT	9	-	+	-
neWAS04p 34	4-b	/	/	1	GRGNPPAYEPQH	12	2-28	4	MQALQTPKLT	10	-	-	-
neWAS04p 35 #	3-30-3	1-26	2	4	GRGGSYYHFDY	11	2-29	4	LQSIQLPLT	9			
neWAS04p 38	3-30	6-19	3	3	VRIAVAAYDAFDI	13	1-12	4	QQANSFPPT	9	-	-	-
neWAS04p 42 #	3-15	5-24	1	2	VGRWLQLRPNWYFDL	15	3-20	2	QQYGSSPPFT	10			
neWAS04p 43	4-30-2	/	/	3	VGVTGAKAFDI	11	1-33	4	QQYDNLPLT	9	-	+	-
neWAS04p 44	3-30-3	1-26	3	4	EYVSIGVGATSYFDY	15	1-39	4	QQSYSTLLT	9	-	-	-
neWAS04p 10	4-59	/	/	6	AYIVADYGGMDV	13							
neWAS04p 11	4-34	2-15	2	3	GGPYCSGGSCYSGSGHTDAFDI	22							
neWAS04p 13	1-2	3-22	2	5	VLKGYDDSSGPNWFDP	17							
neWAS04p 23	1-46	3-22	3	5	ASMIGGGRNWFDP	13							
neWAS04p 24	3-11	3-9	2	4	TYDDRKKISTTDY	14							
neWAS04p 25	3-23	4-17	2	4	DLSYGDYFDY	11							
neWAS04p 08							1-17	4	LQHNSYPLT	9			
neWAS04p 37							3-15	1	QQYNNWRRRT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
neWAS04p 02 #	1-18	1-26	2	5	DGGGSYYF	8	2-23	2	CSYAGSSTPVV	11			
neWAS04p 03 #	4-39	3-22	2	3	DLGSFGAYDSSGYYPGDAFDI	22	1-40	3	QSYDSSLGSGV	11			
neWAS04p 05	3-9	4-17	2	3	AHDYGDYGETGVVAFDI	17	2-23	2	CSYAGSHVV	9	-	-	-
neWAS04p 06	3-15	4-23	2	4	ARIPYGGNRQGSYD	14	1-51	3	GTWDSLSAWV	11	+	+	-
neWAS04p 07	4-30-2	3-16	1	2	VGGLRPRGLYWYFDL	15	3-21	2	QVWDSDDHVV	11	+	-	-
neWAS04p 12	3-30	4-17	3	4	TTVTKSFYD	9	3-21	1	QVWDSDDHYV	11	-	-	-
neWAS04p 15 #	3-66	6-13	3	6	DQVWAAAGPRPYYYYGGMDV	20	1-51	1	GTWDSLSAYV	11			
neWAS04p 17 #	4-30-4	3-10	3	4	ARVVRGVIPIFDY	12	1-44	2	AAWDDSLNGYVV	12			
neWAS04p 18 #	3-21	/	/	2	DLRATTLGWYFDL	13	2-11	3	CSYAGSYTWV	10			
neWAS04p 28	3-64	4-17	2	3	KGRLNGDYESCAFDI	15	2-14	2	SSYTSSSTLEV	11	-	-	-
neWAS04p 36	3-49	5-5	2	2	VQGYSGLYWYFDL	14	3-21	3	QVWDSDDL	10	-	+	-
neWAS04p 40	3-11	6-19	3	4	LVAVAGDFDY	10	3-21	2	QVWDSDDHPVV	12	-	-	-
neWAS04p 41 #	3-30	3-22	2	5	DRLTYDDSSGSLSG	15	3-21	2	QVWDSRGV	8			
neWAS04p 46	3-30-3	5-24	1	3	AWRWLQFGGELDI	13	2-14	2	SSYTSSSSV	10	-	-	-
neWAS04p 48	4-61	/	/	4	DPLPYNSGFYD	11	2-23	2	CSYAGSSTHVV	11	-	-	-
neWAS04p 20							3-21	2	QVWDSDDVV	10			
neWAS04p 31							3-25	2	QSADSSGTVV	10			
neWAS04p 39							1-40	1	QSYDSSLGYYV	11			
neWAS04p 42							3-21	2	QVWDSDDHLV	11			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S16: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 1 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS01p 55 #	3-33	3-3	3	4	DRITIFGVVKKFDY	14	1-17	1	LQHNSYPLA	9			
mnWAS01p 58 #	1-46	3-22	3	4	SWGSVVITDDFDY	13	1-39	1	QQSYSTPWT	9			
mnWAS01p 61	3-30	3-22	2	3	DQAPSSSGYPHDAFDI	16	3-15	4	QQYNNWPLT	9	-	-	-
mnWAS01p 65	3-48	3-22	2	6	DAYYDSSGYNNYYYYGMDV	20	3-11	5	QQRSNWPPIT	10	-	-	-
mnWAS01p 67 #	3-33	4-23	2	4	DLGDYGGNSVVY	12	2-28	1	MQALQTHWT	9			
mnWAS01p 69	3-48	5-5	1	6	DRVWLSGYYYYGMDV	15	1-5	1	QQYNSYPWT	9	+	+	C
mnWAS01p 82	1-46	5-12	3	6	GDIVATIISHYYYYGMDV	18	1-33	2	QQYDNLPLT	9	-	-	-
mnWAS01p 95	1-69	3-9	2	4	GYPNYDILTGYSFDY	15	2-28	1	MQALQTHWT	9	+	+	C
mnWAS01p 71	1-18	3-10	2	6	DLVYYYDSSYYYYYGGMDV	19							
mnWAS01p 78	3-21	3-3	2	6	DHLDFWQRGYYYYGMDV	16							
mnWAS01p 93	3-53	1-20	2	4	GWNWNSRH	9							
mnWAS01p 94	1-24	3-16	1	4	IFRDLRLGELSLYGAPFDY	19							
mnWAS01p 74							1-5	1	QQYNSYWT	8			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS01p 51	3-64	5-24	2	4	GDGYSLDY	8	2-14	2	SSYTSSINVV	10	-	-	-
mnWAS01p 62	3-23	4-17	2	6	FGIDYGDYRGPNNYYYYGMDV	21	3-1	2	QAWDSSTEVV	10	-	-	-
mnWAS01p 63	4-b	2-2	3	5	VGRDIVVPAAIRRNWFDP	20	1-47	3	AAWDDSLSGPV	11	-	-	-
mnWAS01p 68	3-21	2-21	2	3	DDVLDCGGDCYSPDAFDI	18	2-14	2	SSYTSSSTLVV	11	-	+	-
mnWAS01p 70	5-51	6-19	2	4	RSSGWGGNDY	10	3-21	1	QVWDSSTDHYV	11	-	-	-
mnWAS01p 74	4-34	4-17	3	4	GVTVTTRGDFD	11	1-40	3	QSYDSSLGSGV	11	-	-	-
mnWAS01p 75	4-59	7-27	2	5	VSNWGPWFDP	11	1-40	1	QSYDSSLGSGV	11	-	-	-
mnWAS01p 85	1-18	6-13	2	6	ASSWYVPPYYGMDV	15	1-44	3	AAWDDSLNGLV	11	-	-	-
mnWAS01p 88	3-21	3-3	2	4	YNGDDSSGYPRPFDY	16	3-21	1	QVWDSSTDHYV	11	-	-	-
mnWAS01p 90	1-24	3-10	2	6	DLISWTPHGSMDV	14	1-44	3	AAWDDSLNGPV	11	-	-	-
mnWAS01p 92	1-69	3-10	1	6	DSRFGDYYYYGMDV	15	3-21	2	QVWDSSTDHVV	11	-	-	-
mnWAS01p 96	1-69	2-21	3	6	GVVVTATDYYYYGMDV	16	1-51	3	GTWDSLSAWV	11	-	+	-
mnWAS01p 57							3-21	3	QVWDSSTDHWV	11			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S17: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 2 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS02p 50	3-15	4-17	2	6	YGDYDAGGYYYGGMDV	16	3-20	1	QQYGSSPLT	9	-	-	-
mnWAS02p 53	3-23	3-3	3	4	DGRTIFDY	8	3-15	1	QQYNNWPPWT	10	-	-	-
mnWAS02p 56 #	4-34	6-13	3	6	GPPGIAAAGTGDRYYYYGMDV	21	1-9	5	QQLNSYPIT	9			
mnWAS02p 62	4-34	6-13	2	4	GRPVHNSSSWFFFDY	15	4-1	4	QQYYSTPIT	9	-	-	-
mnWAS02p 64	1-24	/	/	3	DYRLYAFDI	9	3-20	2	QQYGSSRT	8	-	-	-
mnWAS02p 65 #	3-23	3-10	2	4	QYYYDSSSYPLSDY	14	3-11	1	QQRINWPRT	9			
mnWAS02p 68	3-30	1-26	2	4	GGSGSYRPLGDY	12	1-33	2	QQYDNLIMYT	10	-	-	-
mnWAS02p 71	1-8	4-17	2	5	VTPDGDGRGRVRFWFD	17	1-33	5	QQYDNLIPIT	9	-	-	-
mnWAS02p 72	3-11	2-15	2	4	GYCSGGSCYVGGMYFDY	17	1-39	3	QQSYSTPRT	9	-	-	-
mnWAS02p 74	4-31	/	/	3	DPPDDDAFDI	10	1-39	1	QQSYSTPWT	9	-	-	-
mnWAS02p 87 #	1-24	3-22	2	3	QTSTYYYDSSGYHDAFDI	18	3-11	3	QQRSNWPLIFT	11			
mnWAS02p 88 #	3-11	3-10	2	4	PYGGSGYTERAADY	14	1-12	4	QRVNRYPPT	9			
mnWAS02p 90	3-7	6-13	3	4	GAAVDY	7	4-1	2	QQYYSTPPYT	10	-	-	-
mnWAS02p 93	1-69	6-13	2	6	SSSWFDYGGMDV	13	2-30	1	MQGTHWPQT	9	-	-	-
mnWAS02p 63	3-7	6-13	3	4	TSLAAAGTWGILGY	14							
mnWAS02p 78	1-69	2-2	2	5	DVRLGYCSSTSCPPENWFD	20							
mnWAS02p 95	1-69	5-24	3	3	DPVEMATINAFDI	13							
mnWAS02p 49							1-5	1	QQYNSYSRT	9			
mnWAS02p 59							3-15	5	QQYNNWPPIT	10			
mnWAS02p 92							1-5	4	QQYNSYPLT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS02p 51	3-21	3-3	3	6	RGVGDYYYGMDV	12	3-25	1	QSADSSGTYV	10	-	-	-
mnWAS02p 54	1-2	1-26	3	1	AIVGATSEYFQH	12	3-21	1	QVWDSSSDHYV	11	-	-	-
mnWAS02p 58	4-39	3-3	2	5	HAYYDFWSGYYIPGWFD	18	3-9	3	QVWDSSSTAWV	10	+	+	C
mnWAS02p 59	3-23	6-13	1	5	QQLVSRWFD	10	3-14	2	SSYSSSTLVV	11	-	-	-
mnWAS02p 69	1-2	3-22	2	4	DEYYDSSGYNY	13	2-14	3	SSYSSSTLGV	11	-	+	-
mnWAS02p 75	3-7	3-10	2	3	DNYGSGSYHNSDAFDI	16	2-14	3	SSYSSSTLV	10	-	-	-
mnWAS02p 83 #	3-15	5-5	3	2	VNPDTAMVFRYFDL	14	2-14	3	SSYSSSTL	9			
mnWAS02p 85	1-69	2-2	3	6	VGDPVVPAAFDYGGMDV	22	2-8	2	SSYAGSNNLV	10	+	+	C
mnWAS02p 91	4-39	4-17	2	5	VYGDYDDWFD	11	1-40	1	QSYDSSLSGLYV	12	-	-	-

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S18: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 3 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	V _k	J _k	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS03p 49 #	1-69	2-21	3	2	SVVVTAIHSYWYFDL	15	3-20	2	QQYGSSSYT	9			
mnWAS03p 50	3-23	/	/	4	WRRSYFDY	8	1-17	4	LQHNSYPLT	9	+	-	-
mnWAS03p 51	4-34	3-10	2	4	GNRGWRGSGSDYFDY	15	3-11	3	HQRSNWPPPT	10	-	-	-
mnWAS03p 52	3-30	6-6	2	4	QSGESSSEVWRYFDY	15	1-39	1	QRSYSTPRT	9	-	-	-
mnWAS03p 54 #	3-9	3-22	3	3	DFGLGVITDDVAFDI	15	1-39	1	QSYSTPWT	9			
mnWAS03p 55	4-59	3-3	2	4	TKTDFWSGYPPSSPYFDY	18	4-1	1	QYYSTPWT	9	+	+	-
mnWAS03p 56 #	3-30-3	4-17	2	3	PRDYGDYDHGLLDAFDI	17	2-28	4	MQALQTPLT	9			
mnWAS03p 58	3-30	4-17	2	4	GYGDYFDY	8	3-11	1	QQRSNWPSWT	10	-	-	-
mnWAS03p 61	4-34	6-6	1	4	KGEQLSFDY	9	3-15	2	QQYNNWPYT	9	-	-	-
mnWAS03p 63	4-4	6-13	2	1	DGPSTSWYNYFQH	13	1-9	2	QQLNSYPRT	9	-	+	-
mnWAS03p 67	3-33	6-19	2	4	DRSGWYFY	8	3-15	4	QRYNNWPPLT	10	-	-	-
mnWAS03p 71	4-34	5-5	3	4	VPLLGDTAMGLDY	13	1-16	3	QQYNSYPRT	9	-	-	-
mnWAS03p 73	1-46	5-5	2	5	DRVEYSYGPFYD	12	3-20	2	QQYGSSSYT	9	-	-	-
mnWAS03p 74	4-34	6-19	3	4	VAGSTDGKGIAGVATYFDY	19	3-11	4	QQRSNWPLT	9	-	-	-
mnWAS03p 75	3-15	7-27	1	2	ARELGHWYFDL	12	1-39	2	QSYSTPYT	9	-	-	-
mnWAS03p 77	4-4	3-9	1	4	NRVRYFGGKGRETLDY	17	1-33	3	QQYDNLPH	9	-	+	N
mnWAS03p 79	3-30-3	3-3	2	2	EGPYDFWSGYFFFRYGYFDL	20	4-1	1	QYYSTPT	8	-	-	-
mnWAS03p 81	1-f	5-5	2	4	PGTGYSYGLVAYHFDY	16	2-28	1	MQALQTPPT	9	-	-	-
mnWAS03p 85	4-34	6-19	2	4	ARSRGWYGFYFDY	14	3-15	4	QQYNNWVLT	9	+	+	C
mnWAS03p 86	4-34	3-10	2	4	GWGGSYSGSSDY	14	1-17	1	LQHNSYPLA	9	-	-	-
mnWAS03p 89 #	1-58	6-13	1	6	DRGIGQQLAVYYYYGMDV	18	1-17	2	LQHNSYPT	9			
mnWAS03p 93 #	1-8	3-22	2	6	ANYDSSGYSSYYYGMDV	19	2-30	2	MQGTHWPPYT	10			
mnWAS03p 53	4-34	6-19	3	4	GIAVAGVFDY	10							
mnWAS03p 80	4-39	6-19	2	4	VSMYSSGWYSLPYFDY	17							
mnWAS03p 83	3-30	3-9	2	3	PQVHDILTGYLKGDAFDI	20							
mnWAS03p 84	1-69	6-6	2	2	SSYSSSARYFDL	13							
mnWAS03p 89	1-58	6-13	1	6	DRGIGQQLAVYYYYGMDV	18							
mnWAS03p 90	3-23	6-19	2	4	GKWPEHSGWDPDF	14							
	VH	D	RF	JH	CDR3(aa)	Length	V _λ	J _λ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS03p 57 #	3-30	3-22	2	6	DGVGGYYDSSGYWERYYYYGMDV	24	2-11	1	CSYAGSYTFV	10			
mnWAS03p 60	3-21	5-5	1	4	EWIQLWLQIDY	11	1-44	3	AAWDDSLNGWV	11	-	-	-
mnWAS03p 66	4-34	6-13	3	3	GSRVAAAGTDAFDI	14	5-37	1	MIWPSNASLYV	11	-	-	-
mnWAS03p 69	3-9	7-27	?	4	PTRSEPGDDTFDY	13	3-21	1	QVWDSSSDYV	10	-	-	-
mnWAS03p 72	3-9	5-5	2	4	DIYSYGSSPDY	11	2-14	1	SSYSSSYV	10	-	-	-
mnWAS03p 87	4-34	2-15	3	5	RPDIVVVAATTGGWFDP	18	2-8	2	SSYAGSNEVV	10	-	-	-
mnWAS03p 88	3-48	6-19	2	5	DSGWSPGWFD	11	3-21	2	QVWDSSSDHVV	11	-	-	-
mnWAS03p 91	4-34	3-10	2	4	GYGSGSYSSYFDY	14	2-11	2	CSYAGSYFV	10	-	-	-
mnWAS03p 92	4-34	/	/	3	GAFDI	6	1-44	3	AAWDDSLNGRV	11	-	-	-
mnWAS03p 95	3-30-3	6-19	3	5	DLLDYSVAGTS	11	1-51	2	GTWDSLSAGRV	12	+	+	-
mnWAS03p 75							1-44	1	AAWDDSLNGYV	11			
mnWAS03p 96							2-23	2	CSYAGSSTVV	10			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers

Table S19: Repertoire and reactivity of antibodies from mature naïve B cells of WAS patient 4 after GT

Ig	HEAVY						LIGHT				REACTIVITY		
	VH	D	RF	JH	CDR3(aa)	Length	Vκ	Jκ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS04p 51	1-69	3-22	2	6	RYYDSSGPDFSHSYYYYGMDV	21	2-28	5	MQALQTPGT	9	-	-	-
mnWAS04p 53	3-9	1-26	2	6	ANHDSGSYSSYYGMDV	17	2-28	1	MQALQTPWT	9	-	-	-
mnWAS04p 57	1-69	1-26	1	5	ATLKWELLEYNWFDP	15	1-33	4	QQYDNLPLT	9	-	-	-
mnWAS04p 58	4-34	5-5	3	4	GRDTAMALYYFDY	13	3-11	2	QQRNSYNT	8	-	-	-
mnWAS04p 59	3-21	/	/	4	PFGQLDNY	8	2-28	2	MQALQTRYT	9	-	-	-
mnWAS04p 60	4-30-4	3-9	2	4	VPNYDILTYTY	11	1-33	5	QQYDNLPLT	9	+	+	C
mnWAS04p 63	1-69	1-26	3	6	IVGATVYYYYGMDV	15	3-11	5	QQRSTWFT	8	-	-	-
mnWAS04p 64	3-30	3-22	2	4	LGADSSGYPPFDY	13	1-17	3	LQHNSYPFT	9	-	-	-
mnWAS04p 65 #	3-30	1-26	2	6	GDSGSYPPFDYYYYYGMDV	19	3-11	4	QQRSNWPLT	9			
mnWAS04p 70 #	4-34	5-5	2	4	VRGYSYGPFDY	11	1-33	1	QQYDNLPLT	9			
mnWAS04p 73	1-69	3-22	2	6	GYDSSGYRAYYYYYGMDV	19	3-20	1	QQYGSPPGT	9	-	-	-
mnWAS04p 75	1-24	6-19	2	3	DLGWGGWHDAFDI	13	3-15	1	QQYNNWPKT	9	-	-	-
mnWAS04p 79	4-30-4	3-16	2	6	CPLPFLGSYGRLSGAEDGMDV	21	1-39	2	QQSYSTPYT	9	-	+	-
mnWAS04p 80 #	3-11	6-19	3	2	DHEPAVAPIFYWYFDL	16	3-11	4	QQRSNWPLT	10			
mnWAS04p 84	1-69	4-17	2	3	DRYRYGAHSAFDI	14	1-33	3	QQYDNLPLT	9	-	-	-
mnWAS04p 85 #	1-18	3-16	2	4	DESYDVVWGSYRYFDY	16	3-11	5	QQRSNWPLIT	11			
mnWAS04p 86 #	3-9	3-22	2	3	DPHYDSDNHDAFDI	14	3-20	2	QQYGSPPSYT	11			
mnWAS04p 88	3-23	3-3	2	4	DYYDFWPPYFDY	12	1-5	2	QQYNSYSSYT	10	-	-	-
mnWAS04p 90	3-30-3	1-7	2	4	VNWNYPAGDYFDY	14	1-33	1	QHQT	5	-	-	-
mnWAS04p 91	5-51	4-4	2	4	PRGSYSNYFDY	12	3-15	2	QQYNNWPEKYT	12	-	-	-
mnWAS04p 49	4-b	6-19	3	5	DLGLAVAGRIVNWFDP	17							
mnWAS04p 54	1-2	6-19	3	4	IADVDPFDY	9							
mnWAS04p 83	1-18	6-6	3	4	SIATIDY	8							
mnWAS04p 56							3-11	1	QQRSNWPPSWT	11			
mnWAS04p 89							3-20	1	QQYGSPLYT	9			
	VH	D	RF	JH	CDR3(aa)	Length	Vλ	Jλ	CDR3 (aa)	Length	Poly	HEp2	Staining
mnWAS04p 55 #	3-9	3-16	1	4	DHLRLGELSSFDY	13	3-25	2	QSADSSGTYVV	11			
mnWAS04p 67	4-61	3-10	3	6	VGTMVRGIPFYGMDV	15	1-36	2	AAWDDSLNGVV	11	-	-	-
mnWAS04p 69 #	3-30	6-19	2	4	LYSSGRFDY	9	3-25	1	QSADSSGTYV	10			
mnWAS04p 71	4-34	/	/	6	GTYYYYYGMVDV	11	1-51	2	GTWDSLSAGV	11	-	-	-
mnWAS04p 72	4-59	3-22	2	6	LSYYDSSGYPPSYYYYYGMDV	22	3-1	2	QAWDSIVV	8	-	+	-
mnWAS04p 73	3-23	3-3	2	5	IEGNYDFWSGYPLENWFDP	19	3-1	1	QAWDSSTGV	9	-	-	-
mnWAS04p 78	3-30	2-2	3	6	STEDIVVPAAPAVHYGMDV	23	1-47	2	AAWDDSLSGAGV	12	-	+	-
mnWAS04p 89	4-b	3-22	1	4	DLWLDYDNYFDY	12	2-23	1	CSYAGSSTYV	10	-	+	C
mnWAS04p 93	4-61	6-13	3	4	TLHTAAAGIHY	11	1-44	2	AAWDDSLNGLV	12	-	-	-
mnWAS04p 96	1-18	3-22	2	3	GSTYDSSGYFPDAFDI	17	2-11	2	CSYAGSYTLV	10	-	-	-
mnWAS04p 66							1-51	2	GTWDSLSAGAV	12			
mnWAS04p 76							3-1	1	QAWDSSTYV	9			

RF, reading frame; #, antibody failed to be expressed; -, non-reactive; +, reactive;

C, diffuse cytoplasmic staining; N, nuclear staining; F, cytoplasmic fibers