

**Table S1.** Immune cell populations, phenotypes and proportions among leukocytes in peripheral blood of each VKH disease patient.

Populations		Model Phenotypes			VKH			
Disease Stage	Patient Number	No.1	No.2	No.3	No.1	No.2	No.3	
Intact live cells (%)		100	100	100	100	100	100	
Lymphocytes	CD3 T cells + B cells + NK cells + plasmablasts	61.1	80.4	46.1	41.5	49.2	9.14	
	CD8 T cells + CD4 T cells + $\gamma\delta$ T cells + MAIT/NKT cells	36.0	39.7	28.5	22.7	33.1	6.48	
	CD3+ CD66b- CD19- CD8+ CD4- CD14- CD161- TCR $\gamma\delta$ - CD123- CD11c-	7.55	3.51	4.98	3.14	4.43	1.56	
	CD8 T cells + CD45RA+ CCR7+ CD27+	1.95	1.51	2.31	0.75	1.68	0.42	
	CD8 T cells + CD45RA- CCR7+ CD27+	0.76	0.43	0.10	0.03	0.04	0.01	
	CD8 T cells + CCR7- CD27+	3.67	0.80	0.76	1.70	1.46	0.43	
	CD8 T cells + CCR7- CD27-	1.18	0.77	1.81	0.66	1.26	0.70	
	CD66b- CD3+ CD8- CD4+ CD14- TCR $\gamma\delta$ - CD11c-	25.7	34.1	22.3	18.2	25.9	4.44	
	CD4 T cells + CD45RA+ CCR7+ CD27+	14.7	25.0	11.9	10.2	13.8	2.12	
	CD4 T cells + CD45RA- CCR7+ CD27+	3.13	4.40	5.14	1.64	2.77	1.17	
	CD4 T cells + CD45RA- CCR7- CD27+	5.85	3.24	4.41	4.20	6.35	0.72	
	CD4 T cells + CD45RA- CCR7- CD27-	2.03	1.30	0.80	2.12	2.95	0.43	
	CD4 T cells + CD25+ CD127- CCR4+	0.73	0.66	0.36	0.12	0.59	0.12	
	CD4 T cells + CXCR3+ CCR6- CXCR5- CCR4-	0.39	0.50	0.46	0.30	1.19	0.24	
	CD4 T cells + CXCR3- CCR6- CXCR5- CCR4+	2.28	2.28	1.94	0.86	1.73	0.21	
	CD4 T cells + CXCR3- CCR6+ CXCR5- CCR4+	3.17	1.24	2.20	2.66	2.19	0.36	
	CD66b- CD3+ CD8dim,- CD4- CD14- TCR $\gamma\delta$ dim,+ CD45RA- CCR7- CD27-	1.40	1.03	0.59	0.41	1.75	0.30	
	MAIT/NKT cells	CD66b- CD3+ CD4- CD14- CD161+ TCR $\gamma\delta$ - CD28+ CD16-	1.28	1.10	0.67	0.98	1.04	0.18
B cells	Naïve	CD3- CD14- CD56- CD16 dim,- CD19+ CD20+ HLA-DR dim,+ CD11c-	12.9	33.0	5.30	7.76	10.3	1.11
	Memory	B cells + CD27-	9.75	28.99	4.71	5.13	9.00	0.88
	Plasmablasts	B cells + CD27+	2.84	3.51	0.57	2.18	1.00	0.20
NK cells	Naïve	CD3- CD14- CD16-,dim CD66b- CD20- CD19+ CD56- CD38++ CD27+	0.28	0.48	0.03	0.45	0.24	0.02
	Early	CD14- CD3- CD123- CD66b- CD45RA+ CD56 dim,+ CD11c-	12.3	7.72	12.3	11.0	5.83	1.56
	Late	NK cells + CD57-	5.97	2.85	1.31	3.79	1.73	0.97
	CD14- CD3- CD123- CD66b- CD45RA+ CD56 dim,+ CD11c-	6.33	4.87	11.0	7.25	4.11	0.59	
Monocytes	Classical	CD3- CD19- CD56- CD66b- HLA-DR+ CD11c+	13.3	8.36	3.40	23.4	24.1	1.55
	Transitional	Monocytes + CD14+ CD38+	11.8	7.77	3.19	22.0	23.3	1.31
	Nonclassical	Monocytes + CD14 dim CD38 dim	0.94	0.44	0.19	1.05	0.73	0.10
Dendritic cells	Monocytes + CD14- CD38- pDCs+ mDCs	0.58	0.15	0.02	0.35	0.07	0.14	
Plasmacytoid DCs	CD3- CD19- CD14- CD20- CD66b- HLA-DR dim,+ CD11c-	0.88	0.82	0.23	0.61	0.70	0.09	
Myeloid DCs	CD3- CD19- CD14- CD20- HLA-DR dim,+ CD11c dim,+ CD123- CD16 dim,- CD38 dim,+ CD294- HLA-D	0.25	0.26	0.05	0.06	0.14	0.00	
Granulocytes	Neutrophils + basophils + eosinophils + CD66b- neutrophils	CD3- CD19- CD14- CD20- HLA-DR dim,+ CD11c-	12.1	4.82	9.81	15.7	13.9	86.1
	CD66b dim,+ CD16+ HLA-DR-	9.31	3.64	7.60	13.4	10.8	85.6	
	HLA-DR- CD66b- CD123 dim,+ CD38+ CD294+	1.89	0.96	0.75	0.9	1.94	0.13	
	CD14- CD3- CD19- HLA-DR- CD294+ CD66b dim,+ CD11c-	0.38	0.11	0.46	0.08	0.22	0.11	
CD66b- neutrophils	CD3- CD19- CD66b- CD56- HLA-DR- CD123- CD45-	0.53	0.11	1.00	1.30	0.88	0.30	

Cell phenotypes are as defined by Bagwell et al [22]. Nomenclature such as dim,+ means that dim to positive events are selected. Bold font denotes classification of leukocytes based on differentiation and function. Italic font denotes the classification based on maturity stage. CD; cluster of differentiation, DCs; dendritic cells, HLA; human leukocyte antigen, MAIT; mucosal associated invariant T,

mDCs; myeloid DCs, NK; natural killer, NKT; natural killer T, pDCs; plasmacytoid DCs, Th; T helper, Tregs; regulatory T cells, VKH; Vogt-Koyanagi-Harada.

**Table S2.** Immune cell populations, phenotypes and proportions among leukocytes in peripheral blood of each healthy control.

Populations		Model Phenotypes		Controls						
Reference Number		No.1	No.2	No.3	No.4	No.5	No.6	No.7		
Intact live cells (%)		100	100	100	100	100	100	100		
Lymphocytes	CD3 T cells + B cells + NK cells + plasmablasts	46.8	45.1	80.8	78.3	61.6	72.8	57.2		
CD3 <sup>+</sup> T cells	CD8 T cells + CD4 T cells + γδ T cells + MAIT/NKT cells	28.6	23.3	50.2	43.3	47.4	43.7	50.6		
CD8 <sup>+</sup> T cells	CD3+ CD66b- CD19- CD8+ CD4- CD14- CD161- TCRγδ- CD123- CD11c-	8.14	8.69	11.3	16.6	14.6	15.0	19.0		
<i>Naïve</i>	CD8 T cells + CD45RA+ CCR7+ CD27+	1.62	5.96	4.92	9.02	7.26	10.77	14.59		
<i>Central memory</i>	CD8 T cells + CD45RA- CCR7+ CD27+	0.10	0.20	0.33	0.24	0.13	0.26	0.11		
<i>Effector memory</i>	CD8 T cells + CCR7- CD27+	1.83	1.57	2.69	3.06	1.60	3.07	1.12		
<i>Terminal effector</i>	CD8 T cells + CCR7- CD27-	4.59	0.97	3.33	4.29	5.61	0.95	3.21		
CD4 <sup>+</sup> T cells	CD66b- CD3+ CD8- CD4+ CD14- TCRγδ- CD11c-	18.4	11.9	32.0	23.2	26.2	22.7	27.7		
<i>Naïve</i>	CD4 T cells + CD45RA+ CCR7+ CD27+	12.9	6.54	18.2	14.2	14.9	12.0	21.1		
<i>Central memory</i>	CD4 T cells + CD45RA- CCR7+ CD27+	2.09	1.95	4.86	1.70	2.62	2.70	1.18		
<i>Effector memory</i>	CD4 T cells + CD45RA- CCR7- CD27+	1.56	1.54	6.19	4.44	0.0	6.95	4.04		
<i>Terminal effector</i>	CD4 T cells + CD45RA- CCR7- CD27-	1.84	1.90	2.77	2.85	8.91	1.02	1.43		
<i>Treg cells</i>	CD4 T cells + CD25+ CD127- CCR4+	0.47	0.30	0.97	0.69	0.44	0.93	0.40		
<i>Th1-like</i>	CD4 T cells + CXCR3+ CCR6- CXCR5- CCR4-	0.04	0.07	1.50	1.00	1.35	1.19	0.52		
<i>Th2-like</i>	CD4 T cells + CXCR3- CCR6- CXCR5- CCR4+	1.33	1.36	1.72	1.19	2.83	1.08	1.69		
<i>Th17-like</i>	CD4 T cells + CXCR3- CCR6+ CXCR5- CCR4+	1.22	1.00	2.06	1.25	0.68	1.43	1.43		
<i>γδ T cells</i>	CD66b- CD3+ CD8dim-, CD4- CD14- TCRγδ dim,+	1.10	2.32	3.16	1.78	6.51	2.27	3.03		
CD4 <sup>-</sup> T Cells										
<i>MAIT/NKT cells</i>	CD66b- CD3+ CD4- CD14- CD161+ TCRγδ- CD28+ CD16-	0.92	0.39	3.80	1.72	0	3.69	0.79		
B cells	CD3- CD14- CD56- CD16 dim,- CD19+ CD20+ HLA-DR dim,+ <i>Naïve</i>	7.52	5.50	10.5	10.7	11.8	13.5	2.05		
<i>Memory</i>	B cells + CD27- <i>Plasmablasts</i>	5.64	4.12	7.40	8.42	9.37	10.30	1.86		
	B cells + CD27+	1.82	1.32	3.04	2.08	2.38	3.11	0.15		
NK cells	CD3- CD14- CD16-dim CD66b- CD20- CD19+ CD56- CD38++ CD27+	0.06	0.06	0.06	0.25	0.07	0.05	0.04		
	CD14- CD3- CD123- CD66b- CD45RA+ CD56 dim,+ <i>Early</i>	10.7	16.2	20.1	24.3	2.43	15.7	4.60		
	NK cells + CD57- <i>Late</i>	2.36	6.13	9.78	5.70	1.52	6.78	1.61		
	NK cells + CD57+	8.31	10.1	11.3	18.6	0.91	8.89	3.00		
Monocytes	CD3- CD19- CD56- CD66b- HLA-DR+ CD11c+	5.78	10.2	6.15	9.88	9.65	11.1	8.42		
	Monocytes + CD14+ CD38+ <i>Classical</i>	4.29	8.59	5.61	9.02	8.55	8.77	6.79		
	Monocytes + CD14 dim CD38 dim <i>Transitional</i>	0.81	1.12	0.43	0.69	0.82	1.69	1.14		
	Monocytes + CD14- CD38- <i>Nonclassical</i>	0.68	0.48	0.11	0.17	0.27	0.63	0.48		
Dendritic cells	pDCs+ mDCs	0.63	0.85	0.79	1.00	0.48	1.08	0.50		
Plasmacytoid DCs	CD3- CD19- CD14- CD20- CD66b- HLA-DR dim,+ CD11c- CD123+	0.12	0.10	0.14	0.35	0.03	0.24	0.05		
Myeloid DCs	CD3- CD19- CD14- CD20- HLA-DR dim,+ CD11c dim,+ CD123- CD16 dim,- CD38 dim,+ CD294- HLA-D	0.51	0.75	0.65	0.65	0.45	0.84	0.46		
Granulocytes	Neutrophils + basophils + eosinophils + CD66b- neutrophils	36.3	31.9	3.41	3.26	17.0	8.23	19.8		
	CD66b dim,+ CD16+ HLA-DR- HLA-DR- CD66b- CD123 dim,+ CD38+ CD294+ CD14- CD3- CD19- HLA-DR- CD294+ CD66b dim,+ <i>Neutrophils</i>	34.3	28.6	1.67	1.60	16.4	5.58	18.7		
	CD14- CD3- CD19- HLA-DR- CD294+ CD66b dim,+ <i>Basophils</i>	0.91	1.46	1.12	1.08	0.04	2.12	0		
	CD14- CD3- CD19- HLA-DR- CD294+ CD66b dim,+ <i>Eosinophils</i>	0.87	1.14	0.01	0.02	0.25	0.04	0.04		
CD66b <sup>-</sup> neutrophils	CD3- CD19- CD66b- CD56- HLA-DR- CD123- CD45-	0.29	0.74	0.60	0.57	0.32	0.49	1.06		

**Table S3.** Hematologic data of No. 1 VKH disease patient.

Acute							
Test Item	Result	Unit	Reference Value	Test Item	Result	Unit	Reference Value
<b>Blood count</b>							
RBC	4.14	x 10 <sup>6</sup> /μL	4.35 ~ 5.55	Endocrine	sIL-2R	U/mL	157 ~ 474
Hb	13.4	g/dL	13.7 ~ 16.8	Immunity	CRP	≤ 0.3 mg/dL	≤ 0.3
Ht	41.6	%	40.0 ~ 50.0	Toxoplasma IgG	< 1.6 IU/mL	< 1.6	
MCV	100.5	fL	83.6 ~ 98.2	Toxoplasma IgM	0.14 S/CO	< 0.83	
MCH	32.4	pg	27.5 ~ 33.2	IgG	1048 mg/dL	870 ~ 1700	
MCHC	32.2	%	31.7 ~ 35.3	IgA	199 mg/dL	110 ~ 410	
RDW	12.8	fL	11.6 ~ 14.0	IgM	36 mg/dL	35 ~ 220	
WBC	6.50	x 10 <sup>3</sup> /μL	3.3 ~ 8.6				
Neutrophil	63.2	%	38.5 ~ 80.5	<b>Infection</b>			
Lymphocyte	28.0	%	16.5 ~ 49.5	RPR	(-)	(-)	
MONO	6.3	%	2.0 ~ 10.0	TPHA	(-)	(-)	
EOSINO	1.9	%	0 ~ 8.5	HBsAg	(-)	(-)	
BASO	0.6	%	0 ~ 2.5	Anti-HCV Ab	0.1	< 0.1	
Platelet	316	x 10 <sup>3</sup> /μL	158 ~ 348	Anti-HIV Ab		(-)	
<b>Biochemistry</b>							
T-Bil	0.50	mg/dL	0.2 ~ 1.2	Anti-HSV Ab	< 4 times	< 4	
AST	21	U/L	8 ~ 30	Anti-VZV Ab	8 times	< 4	
ALT	13	U/L	5 ~ 35	Anti-CMV Ab	< 4 times	< 4	
LDH	191	U/L	100 ~ 225	<b>Coagulation</b>			
Total protein	7.0	mg/dL	6.5 ~ 8.2	PT			
FBS	89	mg/dL	65 ~ 110	Time	10.3 sec	9.9 ~ 11.8	
HbA1c	5.4	%	4.6 ~ 6.2	Activity	108.3 %	80 ~ 127	
BUN	14	mg/dL	8 ~ 20	PT-INR	0.96	0.9 ~ 1.1	
Creatinine	0.77	mg/dL	0.61 ~ 1.13	APTT			
Na	141	mmol/L	135 ~ 147	Time	28.1 sec	24.0 ~ 32.0	
K	4.1	mmol/L	3.5 ~ 5.0	Fibrinogen	339 mg/dL	180 ~ 400	
Cl	102	mmol/L	98 ~ 108	ESR (1 hour)	15 mm	1 ~ 7	
Ca	9.4	mmol/L	8.5 ~ 10.3				
ACE	7.4	IU/L	7.7 ~ 29.4				
eGFR	77.6	mL/min/1.73m <sup>2</sup>					
<b>Remission</b>							
Test item	Result	Unit	Reference value	Test item	Result	Unit	Reference value
<b>Blood count</b>							
RBC	4.14	x 10 <sup>6</sup> /μL	4.35 ~ 5.55	Endocrine	sIL-2R	U/mL	157 ~ 474
Hb	13.4	g/dL	13.7 ~ 16.8	Immunity	CRP	≤ 0.3 mg/dL	≤ 0.3
Ht	41.6	%	40.0 ~ 50.0	Toxoplasma IgG	< 1.6 IU/mL	< 1.6	
MCV	100.5	fL	83.6 ~ 98.2	Toxoplasma IgM	0.14 S/CO	< 0.83	
MCH	32.4	pg	27.5 ~ 33.2	IgG	1048 mg/dL	870 ~ 1700	
MCHC	32.2	%	31.7 ~ 35.3	IgA	199 mg/dL	110 ~ 410	
RDW	12.8	fL	11.6 ~ 14.0	IgM	36 mg/dL	35 ~ 220	
WBC	6.50	x 10 <sup>3</sup> /μL	3.3 ~ 8.6				
Neutrophil	63.2	%	38.5 ~ 80.5	<b>Infection</b>			
Lymphocyte	28.0	%	16.5 ~ 49.5	RPR	(-)	(-)	
MONO	6.3	%	2.0 ~ 10.0	TPHA	(-)	(-)	

EOSINO	1.9	%	0 ~ 8.5	HBsAg	(-)	(-)
BASO	0.6	%	0 ~ 2.5	Anti-HCV Ab	0.1	< 0.1
Platelet	316	x 10 <sup>3</sup> /µL	158 ~ 348	Anti-HIV Ab		(-)
<b>Biochemistry</b>						
T-Bil	0.50	mg/dL	0.2 ~ 1.2	Anti-HSV Ab	< 4	times
AST	21	U/L	8 ~ 30	Anti-VZV Ab	8	times
ALT	13	U/L	5 ~ 35	Anti-CMV Ab	< 4	times
LDH	191	U/L	100 ~ 225	<b>Coagulation</b>		
Total protein	7.0	mg/dL	6.5 ~ 8.2	PT		
FBS	89	mg/dL	65 ~ 110	Time	10.3	sec
HbA1c	5.4	%	4.6 ~ 6.2	Activity	108.3	%
BUN	14	mg/dL	8 ~ 20	PT-INR	0.96	
Creatinine	0.77	mg/dL	0.61 ~ 1.13	<b>APTT</b>		
Na	141	mmol/L	135 ~ 147	Time	28.1	sec
K	4.1	mmol/L	3.5 ~ 5.0	Fibrinogen	339	mg/dL
Cl	102	mmol/L	98 ~ 108	ESR (1 hour)	15	mm
Ca	9.4	mmol/L	8.5 ~ 10.3			
ACE	7.4	IU/L	7.7 ~ 29.4			
eGFR	77.6	mL/min/1.73m <sup>2</sup>				

IgG and IgM levels of toxoplasma are determined by enzyme immunoassay. ACE; angiotensin converting enzyme, ALT; alanine aminotransferase, APTT; activated partial thromboplastin time, AST; aspartate transaminase, BUN; blood urea nitrogen, Ca; calcium, Cl; chlorine, CMV; cytomegalovirus, CRP; C-reactive protein, eGFR; estimated glomerular filtration rate, ESR; erythrocyte sedimentation rate, FBS; fasting blood sugar, Hb; hemoglobin, HBsAg; hepatitis B surface antigen, HCV; hepatitis C virus, HIV; human immunodeficiency virus, HSV; herpes simplex virus, Ht; hematocrit, Ig; immunoglobulin, K; potassium, LDH; lactate dehydrogenase, Na; sodium, MCH; mean corpuscular hemoglobin, MCHC; mean corpuscular hemoglobin concentration, MCV; mean corpuscular volume, PT; prothrombin time, PT-INR; prothrombin time-international normalized ratio, RPR; rapid plasma reagins, RBC; red blood cell, RDW; red blood cell distribution width, sIL-2R; soluble interleukin-2 receptor, TPHA; treponema pallidum hemagglutination test, VZV; varicella-zoster virus, WBC; white blood cells.

**Table S4.** Hematologic data of No. 2 VKH disease patient.

Acute							
Test item	Result	Unit	Reference value	Test item	Result	Unit	Reference value
<b>Blood count</b>							
RBC	4.29	x 10 <sup>6</sup> /μL	4.35 ~ 5.55	Endocrine	sIL-2R	383	U/mL
Hb	14.2	g/dL	13.7 ~ 16.8	Immunity	CRP	≤ 0.3	mg/dL
Ht	43.2	%	40.0 ~ 50.0	Toxoplasma IgG	(-)	IU/mL	≤ 0.3
MCV	100.7	fL	83.6 ~ 98.2	Toxoplasma IgM	(-)	S/CO	< 1.6
MCH	33.1	pg	27.5 ~ 33.2	IgG	1129	mg/dL	< 0.83
MCHC	32.9	%	31.7 ~ 35.3	IgA	158	mg/dL	870 ~ 1700
RDW	13.2	fL	11.6 ~ 14.0	IgM	80	mg/dL	110 ~ 410
WBC	6.10	x 10 <sup>3</sup> /μL	3.3 ~ 8.6	Infection			35 ~ 220
Neutrophil	55.6	%	38.5 ~ 80.5	RPR	(-)		(-)
Lymphocyte	35.6	%	16.5 ~ 49.5	TPHA	(-)		(-)
MONO	5.7	%	2.0 ~ 10.0	HBsAg	(-)		(-)
EOSINO	2.6	%	0 ~ 8.5	Anti-HCV Ab	0.1		< 0.1
BASO	0.5	%	0 ~ 2.5	Anti-HIV Ab			(-)
Platelet	258	x 10 <sup>3</sup> /μL	158 ~ 348	Anti-HSV Ab	32	times	< 4
<b>Biochemistry</b>							
T-Bil	0.59	mg/dL	0.2 ~ 1.2	Anti-VZV Ab	8	times	< 4
AST	21	U/L	8 ~ 30	Anti-CMV Ab	< 4	times	< 4
ALT	18	U/L	5 ~ 35	Coagulation			
LDH	208	U/L	100 ~ 225	PT			
Total protein	7.3	mg/dL	6.5 ~ 8.2	Time	10.8	sec	9.9 ~ 11.8
FBS	96	mg/dL	65 ~ 110	Activity	97.5	%	80 ~ 127
HbA1c	5.8	%	4.6 ~ 6.2	PT-INR	1.01		0.9 ~ 1.1
BUN	9	mg/dL	8 ~ 20	APTT			
Creatinine	0.66	mg/dL	0.61 ~ 1.13	Time	30.9	sec	24.0 ~ 32.0
Na	143	mmol/L	135 ~ 147	Fibrinogen	404	mg/dL	180 ~ 400
K	3.9	mmol/L	3.5 ~ 5.0	ESR (1 hour)	32	mm	1 ~ 7
Cl	106	mmol/L	98 ~ 108				
Ca	9.4	mmol/L	8.5 ~ 10.3				
CK	77	U/L	≤160				
ACE	12.6	IU/L	7.7 ~ 29.4				
eGFR	71.9	mL/min/1.73m <sup>2</sup>					
<b>Remission</b>							
Test item	Result	Unit	Reference value	Test item	Result	Unit	Reference value
<b>Blood count</b>							
RBC	4.15	x 10 <sup>6</sup> /μL	4.35 ~ 5.55	Immunity	CRP	0.7	mg/dL
Hb	13.9	g/dL	13.7 ~ 16.8				≤ 0.3
Ht	42.5	%	40.0 ~ 50.0				
MCV	102.4	fL	83.6 ~ 98.2				
MCH	33.5	pg	27.5 ~ 33.2				
MCHC	32.7	%	31.7 ~ 35.3				
RDW	12.6	fL	11.6 ~ 14.0				
WBC	5.00	x 10 <sup>3</sup> /μL	3.3 ~ 8.6				
Neutrophil	61.6	%	38.5 ~ 80.5				
Lymphocyte	25	%	16.5 ~ 49.5				

MONO	11.6	%	2.0 ~ 10.0
EOSINO	1.6	%	0 ~ 8.5
BASO	0.2	%	0 ~ 2.5
Platelet	234	x 10 <sup>3</sup> /µL	158 ~ 348
<b>Biochemistry</b>			
T-Bil	0.30	mg/dL	0.2 ~ 1.2
AST	24	U/L	8 ~ 30
LDH	228	U/L	100 ~ 225
ALP	69	U/L	38 ~ 113
Total protein	6.3	mg/dL	6.5 ~ 8.2
BUN	23	mg/dL	8 ~ 20
Creatinine	0.82	mg/dL	0.61 ~ 1.13
Na	142	mmol/L	135 ~ 147
K	3.6	mmol/L	3.5 ~ 5.0
Cl	106	mmol/L	98 ~ 108
Ca	9.4	mmol/L	8.5 ~ 10.3

CK; creatine kinase.

**Table S5.** Hematologic data of No. 3 VKH disease patient.

Acute							
Test item	Result	Unit	Reference value	Test item	Result	Unit	Reference value
<b>Blood count</b>							
RBC	4.42	x 10 <sup>6</sup> /µL	4.35 ~ 5.55	CRP	≤ 0.3	mg/dL	≤ 0.3
Hb	13.1	g/dL	13.7 ~ 16.8	Toxoplasma IgG	< 1.6	IU/mL	< 1.6
Ht	40.3	%	40.0 ~ 50.0	Toxoplasma IgM	0.14	S/CO	< 0.83
MCV	91.2	fL	83.6 ~ 98.2	IgG	1256	mg/dL	870 ~ 1700
MCH	29.6	pg	27.5 ~ 33.2	IgA	519	mg/dL	110 ~ 410
MCHC	32.5	%	31.7 ~ 35.3	IgM	52	mg/dL	35 ~ 220
RDW	12.2	fL	11.6 ~ 14.0	<b>Infection</b>			
WBC	5.50	x 10 <sup>3</sup> /µL	3.3 ~ 8.6	RPR	(-)		(-)
Neutrophil	60.6	%	38.5 ~ 80.5	TPHA	(-)		(-)
Lymphocyte	32.4	%	16.5 ~ 49.5	HBsAg	(-)		(-)
MONO	5.2	%	2.0 ~ 10.0	Anti-HCV Ab	0.1		< 0.1
EOSINO	1.4	%	0 ~ 8.5	Anti-HIV Ab			(-)
BASO	0.4	%	0 ~ 2.5	Anti-HSV Ab	< 4	times	< 4
Platelet	272	x 10 <sup>3</sup> /µL	158 ~ 348	Anti-VZV Ab	8	times	< 4
<b>Biochemistry</b>							
T-Bil	1.41	mg/dL	0.2 ~ 1.2	Anti-CMV Ab	8	times	< 4
AST	26	U/L	8 ~ 30	<b>Coagulation</b>			
ALT	21	U/L	5 ~ 35	PT			
LDH	221	U/L	100 ~ 225	Time	10.1	sec	9.9 ~ 11.8
Total protein	7.8	mg/dL	6.5 ~ 8.2	Activity	103.0	%	80 ~ 127
FBS	93	mg/dL	65 ~ 110	PT-INR	0.99		0.9 ~ 1.1
HbA1c	5.9	%	4.6 ~ 6.2	<b>APTT</b>			
BUN	18	mg/dL	8 ~ 20	Time	28.1	sec	24.0 ~ 32.0
Creatinine	0.95	mg/dL	0.61 ~ 1.13	Fibrinogen	310	mg/dL	180 ~ 400
Na	143	mmol/L	135 ~ 147	ESR (1 hour)	29	mm	1 ~ 7

K	4.3	mmol/L	3.5 ~ 5.0				
Cl	105	mmol/L	98 ~ 108				
Ca	10.1	mmol/L	8.5 ~ 10.3				
ACE	21	IU/L	7.7 ~ 29.4				
eGFR	43.9	mL/min/1.73m <sup>2</sup>					
Remission							
Test item	Result	Unit	Reference value	Test item	Result	Unit	Reference value
<b>Blood count</b>							
RBC	4.33	x 10 <sup>6</sup> /μL	4.35 ~ 5.55	Endocrine	sIL-2R	U/mL	157 ~ 474
Hb	13.3	g/dL	13.7 ~ 16.8	Immunity	CRP	mg/dL	≤ 0.3
Ht	40	%	40.0 ~ 50.0	Infection	RPR	(-)	(-)
MCV	92.4	fL	83.6 ~ 98.2	TPHA	(-)	(-)	
MCH	30.7	pg	27.5 ~ 33.2	HBsAg	(-)	(-)	
MCHC	33.3	%	31.7 ~ 35.3	Anti-HCV Ab	0.1		< 0.1
RDW	11.9	fL	11.6 ~ 14.0	Anti-HIV Ab	(-)	(-)	
WBC	7.00	x 10 <sup>3</sup> /μL	3.3 ~ 8.6				
Neutrophil	77.5	%	38.5 ~ 80.5	<b>Coagulation</b>			
Lymphocyte	20.2	%	16.5 ~ 49.5	PT			
MONO	1.9	%	2.0 ~ 10.0	Time	sec		9.9 ~ 11.8
EOSINO	0.3	%	0 ~ 8.5	Activity	%		80 ~ 127
BASO	0.1	%	0 ~ 2.5	PT-INR			0.9 ~ 1.1
Platelet	239	x 10 <sup>3</sup> /μL	158 ~ 348				
<b>Biochemistry</b>							
T-Bil	1.30	mg/dL	0.2 ~ 1.2	APTT			
AST	27	U/L	8 ~ 30	Time	sec		24.0 ~ 32.0
ALT	21	U/L	5 ~ 35				
LDH	276	U/L	100 ~ 225				
FBS	103	mg/dL	65 ~ 110				
HbA1c	6.0	%	4.6 ~ 6.2				
BUN	19	mg/dL	8 ~ 20				
Creatinine	0.93	mg/dL	0.61 ~ 1.13				
Na	141	mmol/L	135 ~ 147				
K	4.2	mmol/L	3.5 ~ 5.0				
Cl	106	mmol/L	98 ~ 108				
eGFR	44.8	mL/min/1.73m <sup>2</sup>					

**Table S6.** Serum cytokine levels in each VKH disease patient.

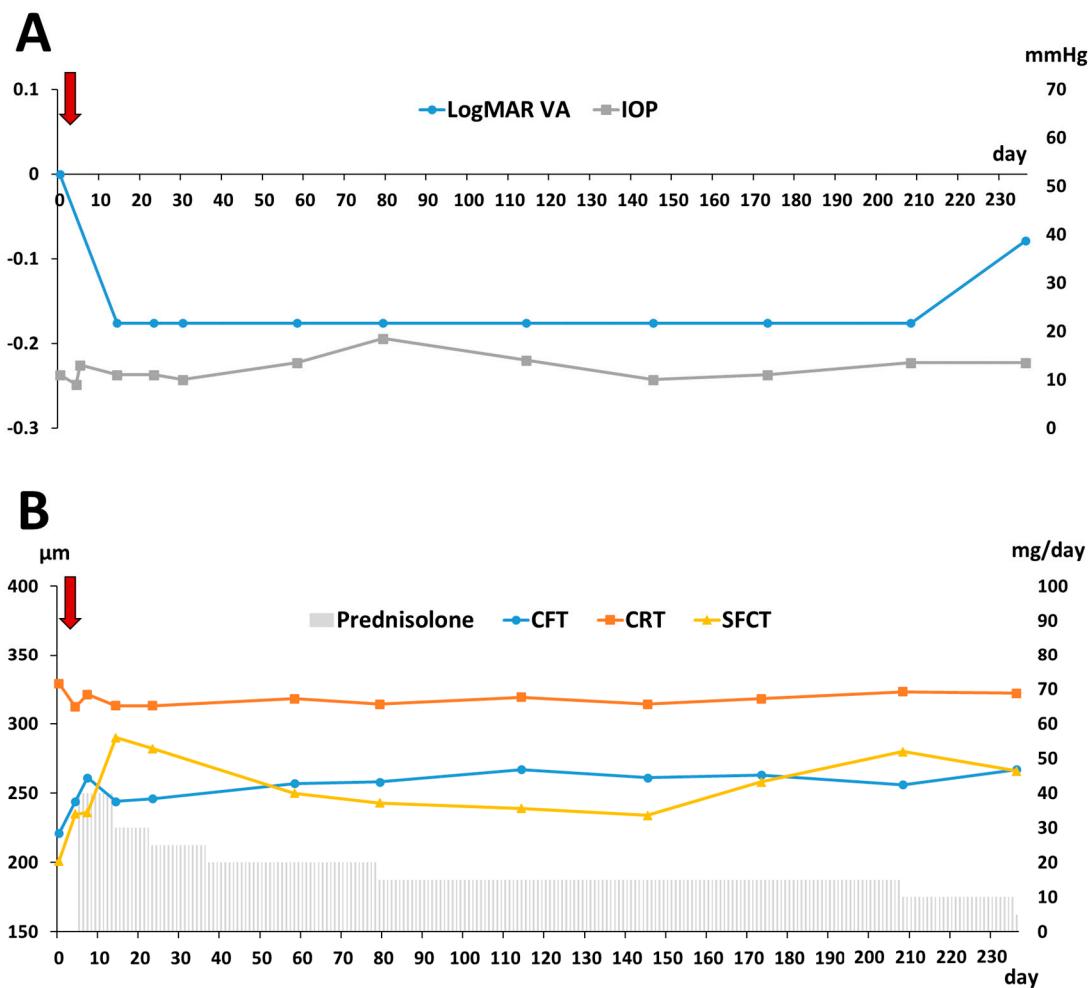
Category	VKH						Detection range		
	Disease stage	Acute			Remission				
		Patient number	No. 1	No. 2	No. 3	No. 1	No. 2	No. 3	
PDGF-BB		1255.7	118.9	345.4		1232.0	54.0	288.4	7.67 to 42619.2
IL-1 $\beta$		0.54	0.39	0		0	0	0	0.28 to 5036.8
IL-1ra		24.0	21.6	36.1		31.2	42.1	36.1	8.56 to 37276.7
IL-2		0	0	0		0	0	0	1.50 to 8778.3
IL-4		1.63	1.22	4.16		3.46	3.44	4.69	0.16 to 3539.9
IL-5		0	0	0		0	0	0	4.02 to 85349.0
IL-6		0	0	0		0	0	0	0.37 to 5969.6
IL-7		33.7	51.7	0		0	25.6	0	1.84 to 36229.8
IL-8		0	0	2.55		1.30	1.50	2.82	0.51 to 10416.3
IL-9		27.4	20.9	49.1		44.4	47.2	43.2	0.92 to 21827.0
IL-10		0	39.8	0		0	18.4	0	0.84 to 12755.7
IL-12		0	24.9	0		0	0	0	1.58 to 21263.1

IL-13	0	4.36	0	0	0	0	0.75	to	5003.2
IL-15	0	0	0	0	0	0	####	to	76422.6
IL-17A	5.20	3.38	0	0	3.05	4.21	2.65	to	35294.2
Eotaxin	139.6	98.4	196.6	168.8	19.4	191.5	0.09	to	1487.0
bFGF	5.99	4.94	0	0	0	0	3.52	to	5445.8
G-CSF	0	519.8	142.8	99.3	362.0	166.5	55.0	to	70102.6
GM-CSF	0	0	0	0	0	0	0.33	to	1822.9
IFN-γ	0	0	0	0	0	1.75	0.74	to	22825.9
IP-10	542.8	847.2	153.0	52.3	70.5	133.0	1.53	to	23765.1
MCP-1	5.10	4.25	8.45	8.78	7.16	5.80	0.34	to	5761.9
MIP-1α	0.19	0	1.18	0.53	0.45	2.37	0.05	to	348.6
MIP-1β	27.1	22.6	26.3	18.8	23.0	36.6	0.46	to	2126.1
RANTES	1010.2	937.3	748.6	649.4	210.7	580.3	1.00	to	5544.2
TNFα	15.5	0	7.32	21.1	0	13.4	2.73	to	53796.5
VEGF-A	0	0	0	0	0	0	19.5	to	69174.0

bFGF; basic fibroblast growth factor, G-CSF; granulocyte colony-stimulating factor, GM-CSF; granulocyte macrophage colony-stimulating factor, IFN; interferon, IP-10; interferon gamma-induced protein 10, IL; interleukin, MIP; macrophage inflammatory protein, MCP; monocyte chemotactic protein, PDGF; platelet derived growth factor, ra; receptor antagonist, RANTES; regulated on activation, normal T-cell expressed and secreted, TNF; tumor necrosis factor, VEGF; vascular endothelial growth factor.

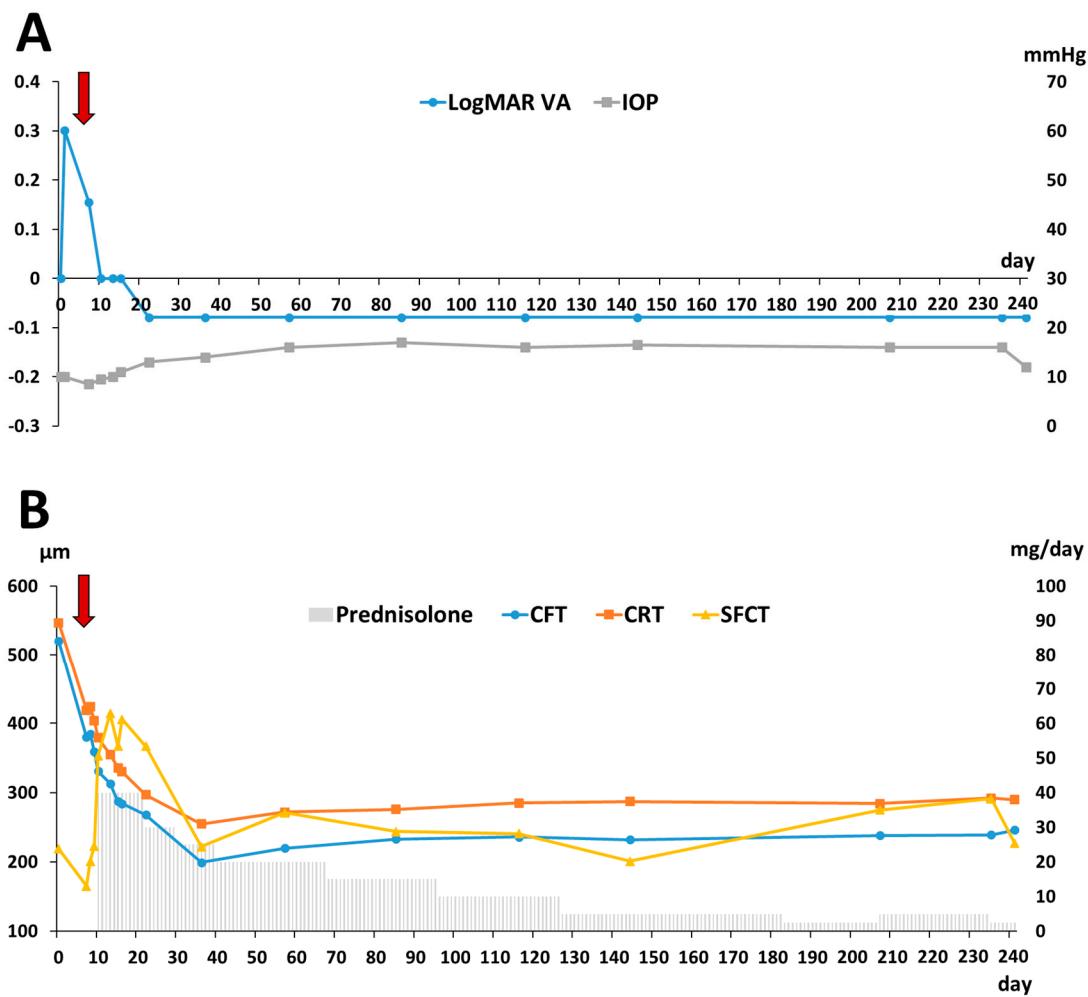
**Table S7.** Serum cytokine levels in each control.

Category	Controls							Detection Range	
	Reference Number	No. 1	No. 2	No. 3	No. 4	No. 5	No. 6	No. 7	
PDGF-BB	83.6	36.8	171.9	360.4	1491.9	1113.4	506.8	7.67	to 42619.2
IL-1β	0.31	0	0	1.08	0.42	0.74	0	0.28	to 5036.8
IL-1ra	0	0	0	41.9	13.1	45.3	22.3	8.56	to 37276.7
IL-2	0	0	0	0	0	0	0	1.50	to 8778.3
IL-4	0.27	0	0	2.07	1.36	2.13	2.03	0.16	to 3539.9
IL-5	0	0	0	0	0	0	0	4.02	to 85349.0
IL-6	0	0	0	0	0	0	0	0.37	to 5969.6
IL-7	0	0	0	33.2	18.1	16.9	0	1.84	to 36229.8
IL-8	0	0	0	0	0	17.6	1.10	0.51	to 10416.3
IL-9	0	0	0	45.6	2.89	35.9	45.6	0.92	to 21827.0
IL-10	0	0	0	0	0	0	0	0.84	to 12755.7
IL-12	0	0	0	0	0	0	0	1.58	to 21263.1
IL-13	0	0	0	0	0	0	0	0.75	to 5003.2
IL-15	0	0	0	0	0	0	0	####	to 76422.6
IL-17A	0	0	0	5.92	0	6.97	0	2.65	to 35294.2
Eotaxin	7.60	4.04	4.54	110.5	57.5	139.7	9.29	0.09	to 1487.0
bFGF	4.67	0	0	7.22	4.67	5.21	0	3.52	to 5445.8
G-CSF	0	0	0	0	0	0	111.1	55.0	to 70102.6
GM-CSF	0	0	0	0	0	0	0	0.33	to 1822.9
IFN-γ	0	0	0	0	0	0	0	0.74	to 22825.9
IP-10	48.3	117.7	63.9	778.0	28.5	332.3	53.0	1.53	to 23765.1
MCP-1	0	0	0	3.86	0	0	0.60	0.34	to 5761.9
MIP-1α	0	0	0	0.51	0	0.93	1.62	0.05	to 348.6
MIP-1β	3.41	0	0	38.7	5.78	34.5	31.1	0.46	to 2126.1
RANTES	28.6	46.9	66.9	179.7	488.1	1446.2	93.5	1.00	to 5544.2
TNFα	0	0	0	15.5	0	31.6	0	2.73	to 53796.5
VEGF-A	0	0	0	0	0	0	0	19.5	to 69174.0

**Figure S1.** Clinical course of No. 1 VKH disease patient under glucocorticoid therapy.

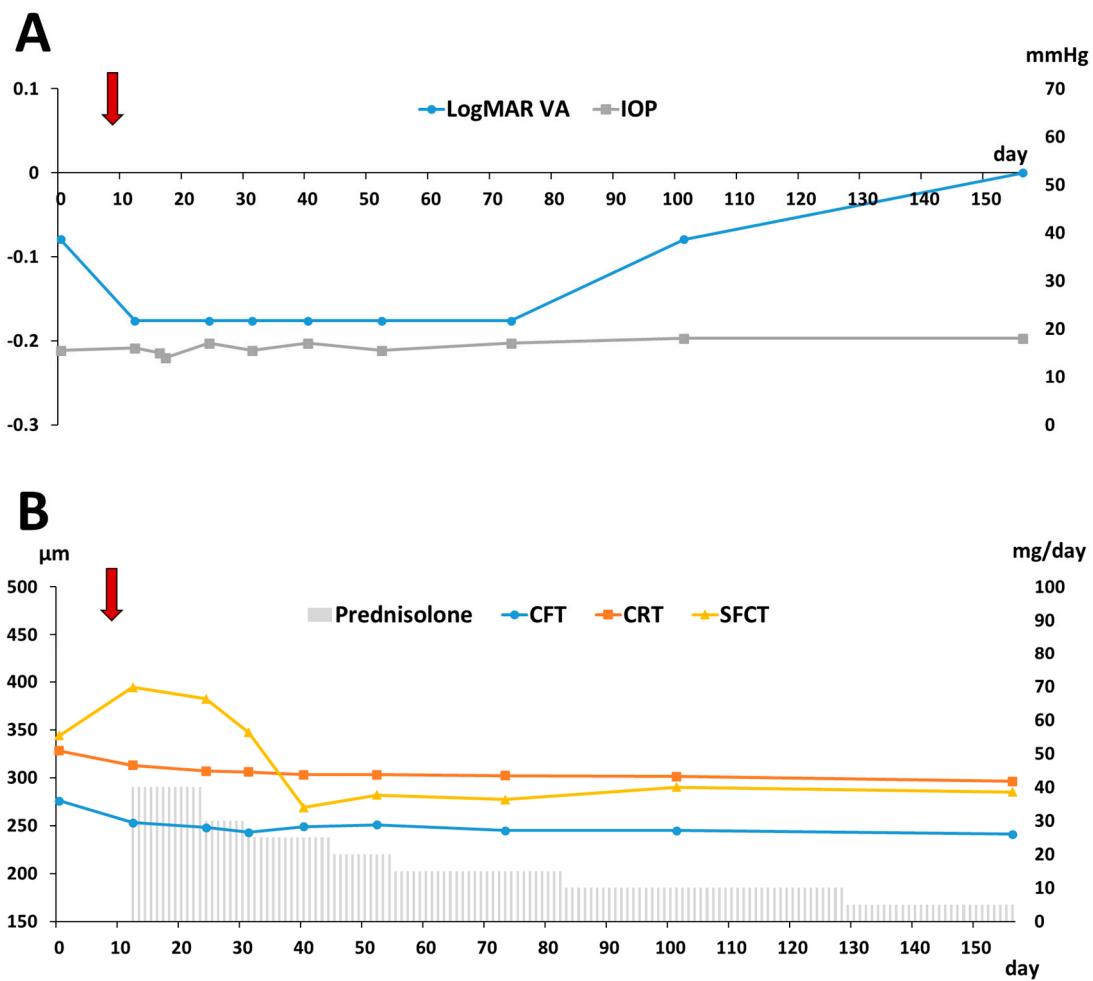
Time courses of (A) logMAR VA, IOP, and (B) dose of prednisolone, CFT, CRT, SFCT in No.1 VKH disease patient under GC therapy are shown. Left vertical axis shows logMAR VA in (A), CFT, CRT, SFCT in (B), and right vertical axis means IOP in (A), dose of prednisolone in (B). Horizontal axis denotes number of days passed since first medical examination. Red arrows in (A, B) indicate intravenous pulses of methylprednisolone (1,000 mg/day for 3 days). CFT; central foveal thickness, CRT; central retinal thickness, GC; glucocorticoid, SFCT; subfoveal choroidal thickness. VKH; Vogt-Ko yanagi-Harada.

**Figure S2.** Clinical course of No. 2 VKH disease patient under glucocorticoid therapy.



Time courses of (A) logMAR VA, IOP, and (B) dose of prednisolone, CFT, CRT, SFCT in No.2 VKH disease patient under GC therapy are presented.

**Figure S3.** Clinical course of No. 3 VKH disease patient under glucocorticoid therapy.



Time courses of (A) logMAR VA, IOP, and (B) dose of prednisolone, CFT, CRT, SFCT in No.3 VKH disease patient under GC therapy are shown.