

Supplementary Table 1. Baseline characteristics

Patient	Age	Sex	Previous Therapies (No.)	Etiology	Fever	Splenomegaly	Hgb (g/dL)	ANC (K/uL)	Plt (K/uL)	Trig (mg/dL)	Fibr (mg/dL)	Hemophagocytosis	Ferritin (mg/dL)	Low/absent NK cell activity	sIL-2R (unit/mL)
1	29	F	1 (corticosteroids)	SLE, IE, Primary	Yes	Yes	8.3	0	23	381	111	Yes	1867.8	Yes	78,200
2	36	M	1 (corticosteroids)	Idiopathic	Yes	Yes	7.5	1.9	107	229	480	Yes	11,509.1	Yes	1,216
3	33	F	2 (corticosteroids, Anakinra)	AOSD	Yes	Yes	6.2	1.1	26	223	154	Yes	16,500	Yes	4,242
4	43	F	0	CMV	Yes	Yes	7.0	0.8	68	311	90	No	5,607.9	Yes	14,550
5	40	F	0	Idiopathic	Yes	Yes	7.8	2.3	169	203	401	Yes	5,102.6	N/A	3,460
6	50	M	0	Idiopathic	Yes	Yes	11.0	8.4	155	354	251	Yes	>16,500	N/A	14,588
7*	62	M	5 (IVIg, corticosteroids, HLH-94, tacrolimus, tocilizumab)	Idiopathic	Yes	Yes	6.5	1.3	15	296	576	Yes	14,629	N/A	4,787
8*	41	F	3 (IVIg, corticosteroids, HLH-94)	Idiopathic	Yes	No	6.1	1.8	32	508	192	Yes	170,403	N/A	10,028
9*	21	M	4 (tocilizumab, corticosteroids, HLH-94, anakinra)	AOSD	Yes	No	5.9	10.3	55	718	162	Yes	520,761	N/A	3,467
10*	56	F	1 (Corticosteroids)	SLE; Disseminated Candidiasis	Yes	Yes	7.0	<1.0	18	314	187	Yes	25,315	Yes	N/A
11*	70	F	0	Hodgkin Lymphoma; EBV viremia	Yes	No	7.0	2.5	33	703	589	Yes	31,774	N/A	12,597
12*	73	M	0	Idiopathic	Yes	Yes	6.0	<0.3	12	317	163	Yes	14,601.7	N/A	12,110
13*	30	M	1 (HLH-94 and rituximab)	CAEBV	Yes	Yes	7.1	0.6	7	423	167	N/A	363.3	N/A	13,018

*Patients treated "off study".

Abbreviations: Absolute neutrophil count (ANC), Adult onset Still's disease (AOSD), Chronic active EBV viremia (CAEBV), hemoglobin (Hgb), platelets (Plt), systemic lupus erythematosus (SLE), triglycerides (trig), infective endocarditis (IE), fibrinogen (fibr), not assessed (N/A).