

Table S1: Cytokine levels (pg/ml) from the patient's plasma and samples from five healthy individuals. ND: not detected.

Cytokines (pg/ml)	First measurements		Second measurements	
	Healthy Individuals	Patient	Healthy Individuals	Patient
		During Flares		Immediately following flares
IL-1 β	ND ND ND ND ND	202.04	ND ND ND ND ND	50
IL-6	ND ND ND ND ND	147.92	ND ND ND ND ND	41
IL-18	378 346 154 692 1946	3332.59	631 595 1382 416 336	5262
TNF α	ND ND ND ND ND	185.27	ND ND ND ND ND	2.16

Table S2: Quantitative measurements of the NF- κ B luciferase assay

	Without NAIP				With NAIP			
pEV	0.1	0.08	0.09	0.08	0.08	0.08	0.06	0.07
pNLRC4_WT	0.5	0.39	0.31	0.25	1.47	1.18	1.21	0.94
pNLRC4_Thr133Ile	0.72	0.73	0.67	0.61	2.43	3.45	3.84	2.59
pNLRC4_Ser171Phe	1.26	1.16	1.16	0.99	4.61	4.48	3.45	3.75
pNLRC4_Thr177Ala	0.63	0.56	0.63	0.46	3.15	2.75	3.39	3.24