

SUPPLEMENTAL MATERIAL

Rodero et al., <https://doi.org/10.1084/jem.20161451>

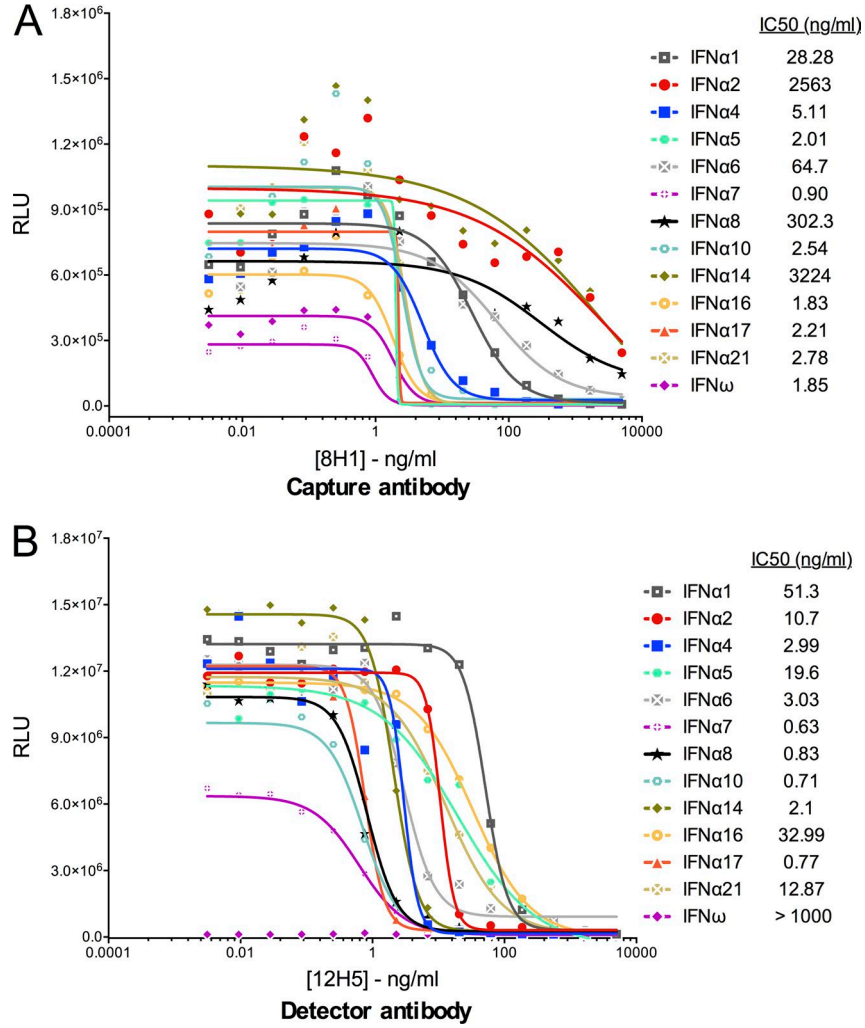


Figure S1. **IC50 values of mAbs used in Simoa assay for all IFNα subtype.** 10,000 HEK 293 MSR cells were seeded in white half-area 96-well plates (Corning) and reverse-transfected with 50 ng premixed ISRE-Firefly luciferase reporter and Renilla luciferase constructs (QIAGEN) using FuGENE HD according to the manufacturer's instructions (Promega). The Renilla luciferase-expressing construct served as an internal normalization control. Cells were incubated overnight in Opti-MEM I Reduced Serum Medium supplemented with 0.1 mM nonessential amino acids, 1 mM sodium pyruvate, 0.5% fetal bovine serum (Thermo Fisher Scientific) at 37°C, 5% CO<sub>2</sub> in a humidified atmosphere. After overnight incubation, cells were stimulated for 24 h with medium containing mixtures of recombinant human IFNα with or without anti-IFNα mAbs (A: 8H1 Ab, B: 12H5 Ab) or control IgG that had been preincubated for 1 h at 37°C. After 24 h of stimulation, dual luciferase reporter assays were performed according to the manufacturer's instructions (Promega). RLU, relative luciferase units.

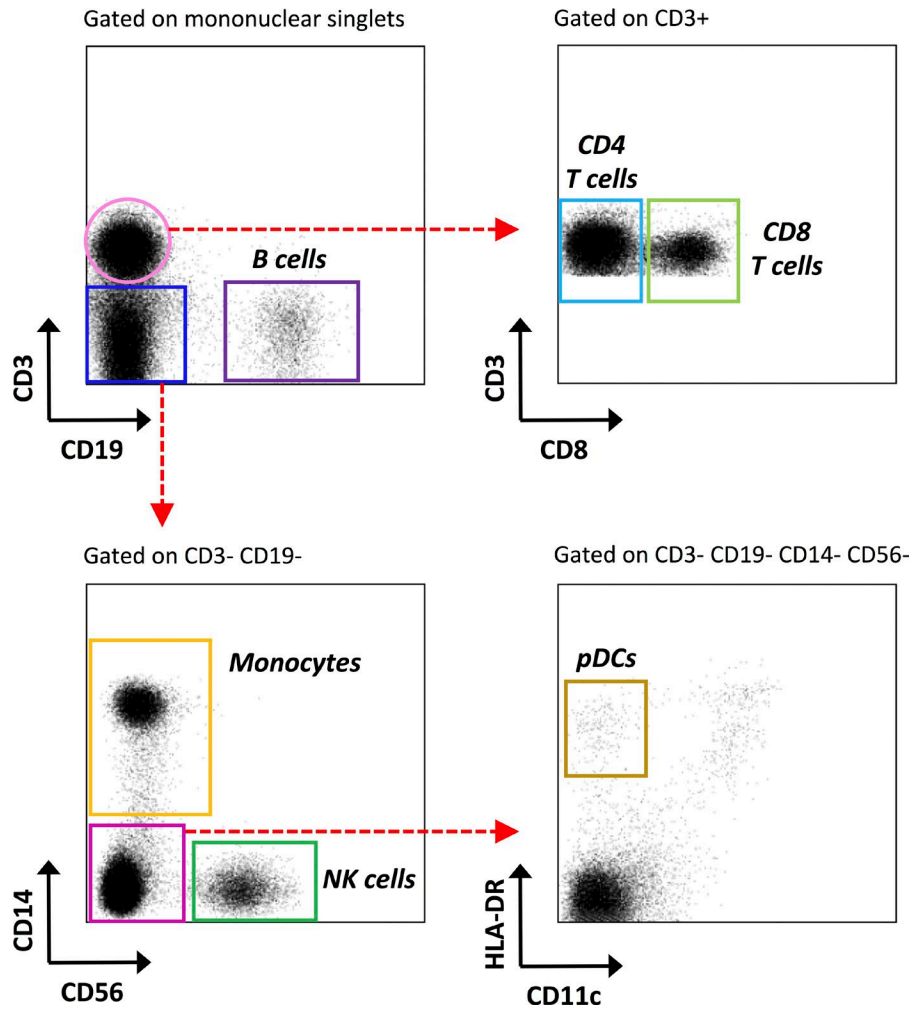


Figure S2. **Flow cytometry gating strategy.** Flow cytometry gating strategy for the isolation of CD8<sup>+</sup> and CD4<sup>+</sup> T cells, B cells, NK cells, monocytes, and pDCs.

Table S1. Retinal vasculopathy with cerebral leukodystrophy

Patient	Sex	Age at sampling <i>yr</i>	Gene	ISG score	IFN activity	IFN $\alpha$ serum <i>fg/ml</i>	IFN $\alpha$ CSF <i>fg/ml</i>
RVCLM001	M	54	TREX1	ND	<2	0.69	ND
RVCLM003	M	23	TREX1	ND	<2	1.81	0.69
RVCLM004	F	62	TREX1	1.4	<2	24.31	ND
RVCLM005	F	52	TREX1	ND	<2	1.12	0.69
RVCLM006	F	61	TREX1	ND	<2	1.10	ND
RVCLM007	F	51	TREX1	ND	<2	0.96	ND
RVCLM008	F	53	TREX1	ND	<2	3.60	0.69
RVCLM010	M	57	TREX1	ND	<2	3.49	ND
RVCLM011	F	22	TREX1	ND	<2	1.49	2.40
RVCLM012	M	59	TREX1	2.1	<2	1.21	ND
RVCLM013	F	44	TREX1	ND	<2	0.92	0.69
RVCLM014	F	19	TREX1	ND	<2	2.36	ND
RVCLM015	M	53	TREX1	ND	<2	2.09	0.69
RVCLM016	F	52	TREX1	ND	<2	1.92	0.69
RVCLM017	F	23	TREX1	ND	<2	10.82	ND
RVCLM018	M	25	TREX1	ND	<2	0.69	ND
RVCLM020	F	21	TREX1	ND	<2	0.69	ND
RVCLM021	F	57	TREX1	ND	<2	1.39	ND
RVCLM023	F	31	TREX1	ND	<2	1.32	0.69
RVCLM025	F	62	TREX1	ND	<2	17.26	55.72
RVCLM026	M	36	TREX1	ND	<2	0.69	ND
RVCLM027	F	61	TREX1	ND	<2	0.69	ND
RVCLM029	F	44	TREX1	ND	<2	0.93	0.69
RVCLM034	M	42	TREX1	ND	<2	1.76	0.69
RVCLM036	M	55	TREX1	ND	<2	7.65	ND
RVCLM043	F	64	TREX1	ND	<2	1.18	ND
RVCLM052	M	38	TREX1	ND	<2	1.72	17.95
RVCLM055	F	55	TREX1	ND	<2	11.75	ND
RVCLM062	M	24	TREX1	ND	<2	28.84	ND

F, female; M, male; ND, not determined.

Table S2. JDM

Patient	Sex	Age at sampling	ISG score	IFN activity	IFN $\alpha$	Treatment
		<i>yr</i>			<i>fg/ml</i>	
F745	M	7	8.1	ND	48.5	HCO, MMF, steroids
F857	M	5	25.9	12	196.9	Immunoadsorption ivaquine, steroids
F859	F	7	ND	ND	160.8	Bolus MP, steroids
F875	F	19	ND	ND	8.9	Steroids
F900	F	9	ND	ND	116.5	MMF, steroids
F907	F	12	2.4	ND	1.6	MTX, steroids
F917	F	15	ND	ND	202.8	HCO, MTX, steroids
F918	M	14	8.5	ND	26.7	Steroids
F938	F	13	9.2	ND	94.1	MMF, steroids
F940	F	6	46.7	ND	364.5	MTX
F942	F	11	ND	ND	17.6	MMF
F945	M	10	4.6	ND	153.4	HCO, i.v. Ig, RTX, steroids
F946	F	8	2.7	ND	19.1	HCO, i.v. Ig, steroids
F947	F	15	16.1	<3	3.7	Bolus MP, steroids
F948	M	16	37.1	100	20,149.5	HCO, MMF
F976	M	14	0.4	ND	0.7	Steroids
F1011	F	12	17.7	ND	106.7	Aspirin, HCO, MMF, steroids
F1022	F	14	18.6	50	939.4	—
F1051	F	13	17.8	<3	14.3	MTX, steroids
F1066	M	18	ND	ND	2,274.0	—
F1067	F	11	4.1	ND	33.1	—
F1073	F	13	10.2	ND	92.7	Plasmapheresis
F1091	M	15	12.0	ND	82.2	MTX
F1093	M	11	29.8	ND	212.6	MTX, steroids
F1105	F	14	31.9	12	33.0	Bolus MP, MTX, steroids
F1106	F	12	18.3	ND	7,457.6	—
F1162	F	17	9.0	ND	24.2	HCO, MMF, MTX, steroids
F1175	F	14	43.1	ND	119.8	Steroids
F1177	F	13	ND	ND	95.7	MTX, steroids
F1178	F	15	ND	ND	69.4	MTX, steroids
F1193	M	9	8.5	ND	50.0	HCO, MTX, steroids
F1194	F	13	ND	ND	10.5	AZA, 5-ASA
F1209	F	9	ND	ND	75.7	Steroids
F1245	M	18	ND	ND	55.9	HCO, MTX
F1305	F	9	ND	ND	14.0	—
F1320	M	12	ND	ND	56.1	Steroids
F1326	F	16	ND	ND	0.3	HCO, MTX, steroids
F1124	F	15	ND	ND	1.3	—
F1435	M	10	ND	ND	16.5	—
F1511	F	19	22.0	ND	116.6	—
F1527	F	16	ND	ND	68.4	—
F1540	F	13	ND	ND	7.9	Steroids

5-ASA, 5-aminosalicylic acid; AZA, azathioprine; F, female; HCO, hydroxychloroquine; M, male; MMF, mycophenolate mofetil; MP, methylprednisolone; MTX, methotrexate; ND, not determined; RTX, rituximab.

Table S3. SLE

Patient	Phenotype	Sex	Age at sampling	ISG score	IFN activity	IFN $\alpha$
			yr			fg/ml
LUP001	SLE	F	41	ND	ND	0.69
LUP002	SLE	F	61	ND	ND	189.73
LUP003	SLE	F	53	ND	ND	67.08
LUP004	SLE	F	55	ND	ND	1,029.55
LUP005	SLE	F	59	ND	ND	23.59
LUP006	SLE	F	40	ND	ND	815.33
LUP007	SLE	F	47	ND	ND	0.69
LUP008	SLE	F	75	ND	ND	9.53
LUP009	SLE	F	20	ND	ND	80.55
LUP010	SLE	F	67	ND	ND	0.69
LUP011	SLE	F	57	ND	ND	0.69
LUP012	SLE	F	43	ND	ND	343.41
LUP013	SLE	F	67	ND	ND	0.69
LUP014	SLE	F	51	ND	ND	748.31
LUP015	SLE	F	23	ND	ND	5.37
LUP016	SLE	F	67	ND	ND	2.39
LUP017	SLE	F	34	ND	ND	4.31
LUP018	SLE	F	30	ND	ND	146.68
LUP020	SLE	F	54	ND	ND	7.18
LUP021	SLE	F	70	ND	ND	18.86
LUP022	SLE	F	28	28.6	ND	190.44
LUP023	SLE	F	50	0.47	ND	0.69
LUP024	SLE	F	47	0.74	ND	0.69
LUP025	SLE	F	63	27.32	ND	1,969.67
LUP026	SLE	F	67	16.05	ND	50.96
LUP027	SLE	F	42	22.84	ND	1,204.64
LUP028	SLE	F	43	12.23	ND	31.69
LUP029	SLE	F	40	25.11	ND	0.69
LUP030	SLE	F	67	1.92	ND	0.69
LUP032	SLE	M	60	16.7	ND	217.50
LUP034	SLE	F	38	24.05	ND	580.45
LUP035	SLE	F	41	1.11	ND	0.69
LUP036	SLE	M	32	0.31	ND	0.69
LUP037	SLE	F	41	0.75	ND	0.69
LUP039	SLE	F	48	0.18	ND	0.69
LUP040	SLE	F	50	12.39	ND	49.02
LUP041	SLE	F	ND	ND	ND	0.69
LUP042	SLE	F	45	6.64	ND	9.28
LUP043	SLE	F	66	6.81	ND	15.91
LUP044	SLE	F	24	0.21	ND	0.69
LUP045	SLE	F	54	0.17	ND	0.69
LUP046	SLE	F	52	0.27	ND	83.73
LUP047	SLE	F	30	ND	ND	9.39
LUP048	SLE	F	57	17.9	ND	113.59
LUP049	SLE	F	22	19.23	ND	694.19
LUP050	SLE	F	40	0.26	ND	0.69
LUP051	SLE	M	44	5.01	ND	6.02
CTD002	SLE	F	44	ND	ND	501.50
CTD009	SLE	F	30	ND	ND	292.40
CTD014	SLE	F	40	ND	ND	111.11
CTD017	SLE	F	35	ND	ND	267.97
CTD021	SLE	F	29	ND	ND	1,657.47
CTD025	SLE	F	62	ND	ND	3.10
CTD026	SLE	F	49	ND	ND	328.93
CTD031	SLE	F	32	ND	ND	0.69
CTD033	SLE	F	58	ND	ND	236.83
CTD060	SLE	F	23	ND	ND	0.69
CTD061	SLE	M	17	ND	ND	67.62
CTD064	SLE	F	62	ND	ND	12.11
CTD066	SLE	F	51	ND	ND	75.15
CTD067	SLE	F	52	ND	ND	0.69
CTD073	SLE	F	47	ND	ND	13.23

Table S3. SLE (Continued)

Patient	Phenotype	Sex	Age at sampling	ISG score	IFN activity	IFN $\alpha$
CTD074	SLE	F	45	ND	ND	0.69
CTD075	SLE	F	30	ND	ND	1,035.89
CTD096	SLE	F	51	ND	ND	2,044.52
CTD099	SLE	F	55	ND	ND	0.69
F1017	JSLE	M	15	1.9	ND	11.62
F1054	JSLE	F	11	24	ND	986.27
F1078	JSLE	F	15	11.4	ND	112.26
F898	JSLE	F	12	11.3	ND	226.53
F472	JSLE	F	15	25.9	100	4,379.37
F885	JSLE	F	15	5.4	2	21.40

F, female; M, male; ND, not determined.

Table S4. Monogenic interferonopathies

Patient	Phenotype	Sex	Age at sampling	Gene	ISG score	IFN activity	IFN $\alpha$
			yr				fg/ml
F759	AGS	F	5	<i>ADAR1</i>	18.6	ND	111.20
F788	AGS	F	3	<i>ADAR1</i>	4.6	ND	36.55
F755_1	AGS-like (proband)	M	3	<i>IFIH1</i>	31.9	ND	218.51
F755_2	FCL-like (sibling of proband)	M	4	<i>IFIH1</i>	59.8	300	10,188.83
F755_3	SMS-like (father of F755_1 and F755_2)	M	42	<i>IFIH1</i>	16.7	<2	38.74
F427	AGS	M	6	<i>RNASEH2A</i>	9.7	ND	71.35
F127	AGS	M	9	<i>RNASEH2B</i>	5.2	ND	30.42
F190	AGS	F	10	<i>RNASEH2B</i>	5.4	ND	59.18
F711	AGS	F	4	<i>RNASEH2B</i>	1.3	ND	5.32
F903	AGS	F	4	<i>RNASEH2B</i>	7.4	ND	248.65
F1023	AGS	F	1	<i>RNASEH2B</i>	6.0	ND	2,223.52
F1169	AGS	M	2	<i>RNASEH2B</i>	7.0	ND	306.36
F088_1	AGS (sibling of F088_2)	M	11	<i>RNASEH2C</i>	8.6	ND	310.38
F088_2	AGS (sibling of F088_1)	M	16	<i>RNASEH2C</i>	9.8	ND	506.15
F559	AGS	M	11	<i>SAMHD1</i>	16.2	75	7,699.83
F1119	AGS	F	3	<i>SAMHD1</i>	17.4	4	92.00
F1289	AGS	M	2	<i>SAMHD1</i>	ND	ND	14,236.0 <sup>a</sup>
F769	AGS	M	14	<i>TREX1</i>	16.8	ND	3,379.48
F958	AGS	F	1	<i>TREX1</i>	3.6	ND	1,221.43
F1081_1	FCL (proband)	F	28	<i>TREX1</i>	6.2	ND	175.44
F1081_2	Asymptomatic mother of F1081_1	F	49	<i>TREX1</i>	7.7	ND	56.38
F831	STING	F	5	<i>TMEM173</i>	ND	9	1,627.99
F876	STING	M	9	<i>TMEM173</i>	24.9	ND	14,735.41
F1058	STING	F	8	<i>TMEM173</i>	13.9	ND	817.80
F1182	STING	M	11	<i>TMEM173</i>	24.5	ND	942.90
F1295	STING	M	9	<i>TMEM173</i>	ND	ND	2,416.50
F1429	STING	M	8	<i>TMEM173</i>	21.1	ND	896.30
F1802	STING	M	16	<i>TMEM173</i>	ND	ND	44,579.70

F, female; FCL, familial chilblain lupus; SAVI, STING-associated vasculopathy with onset in infancy; SMS, Singleton–Merten syndrome; M, male; ND, not determined.

<sup>a</sup>Measured in CSF.

Table S5. **CNS inflammation**

Patient	Sex	Age at sampling <i>yr</i>	Phenotype	IFN $\alpha$ activity CSF	IFN $\alpha$ CSF <i>fg/ml</i>
152-86	F	<1	Enteroviral meningitis	25	14,371.2
153-19	M	<1	Enteroviral meningitis	18	3,786.4
166-84	M	<1	Enteroviral meningitis	38	6,432.2
165-86	F	<1	Enteroviral meningitis	38	11,173.4
157-23	M	<1	Enteroviral meningitis	25	4,817.7
163-12	F	59	Herpes zoster meningitis	<2	84.8
160-92	F	26	Herpes zoster meningitis	18	2,870.7
165-77	M	<1	Varicella meningitis	3	3,385.7
152-100	M	5	Viral meningitis <sup>a</sup>	18	5,666.8
165-81	F	46	Herpes simplex encephalitis	9	2,737.6
154-78	F	36	Herpes simplex encephalitis	75	27,615.3
163-52	F	90	Herpes zoster encephalitis	3	1,000.9
160-47	M	76	Herpes zoster encephalitis	6	2,437.4
165-46	M	39	Viral encephalitis <sup>a</sup>	2	408.6
159-3	M	<1	Viral meningoencephalitis <sup>a</sup>	>200	33,035.6
159-75	M	<1	Post-infectious encephalitis	9	8,581.6
162-12	M	2	Post-infectious encephalitis	6	1,711.1
165-42	M	<1	Post-infectious encephalitis	6	4,174.2
158-39	M	14	Post-infectious encephalomyelitis	12	2,0314.3

F, female; M, male.

<sup>a</sup>Diagnosis of probable viral meningitis or encephalitis was established on clinical, biological, and imaging data.Table S6. **Healthy controls**

Individual	IFN $\alpha$ <i>fg/ml</i>
816-19	0.69
816-16	0.69
816-12	0.69
816-08	0.69
816-07	0.69
816-06	0.69
816-05	2.01
816-01	0.82
816-50	0.69
816-47	0.69
816-33	0.69
816-32	0.69
816-24	0.69
816-22	0.69
816-21	13.13
816-20	0.69
816-89	0.69
816-83	0.69
816-79	1.18
816-52	5.99

Table S7. Information on data used for Fig. 3 A

Patients	Phenotypes	Gene	IFN $\alpha$	IFN activity
			<i>fg/ml</i>	
F788	AGS	<i>ADAR1</i>	36.5	3
F755_3	SMS-like	<i>IFIH1</i>	38.7	<2
F755_2	FCL-like	<i>IFIH1</i>	10,188.8	300
AGS1169	AGS	<i>RNASEH2B</i>	379.6	3
AGS1023	AGS	<i>RNASEH2B</i>	2,527.5	12
F559	AGS	<i>SAMHD1</i>	7,699.8	75
F1081_1	FLC	<i>TREX1</i>	175.4	<2
F1081_2	Asymptomatic	<i>TREX1</i>	56.4	<2
F831	SAVI	<i>TMEM173</i>	722.8	9
F876	SAVI	<i>TMEM173</i>	14,735.4	200
AGS1429	SAVI	<i>TMEM173</i>	896.3	12
F1022	JDM		939.4	50
F1051	JDM		14.3	<3
F1066	JDM		2,274.2	25
F1073	JDM		54.1	2
F1093	JDM		9,100.7	50
F1105	JDM		33.0	12
F1105	JDM		13.7	<2
F1106	JDM		6,650.3	75
F1175	JDM		238.1	2
F1175	JDM		119.8	12
F1193	JDM		48.3	<2
F857	JDM		196.9	12
F857	JDM		185.2	2
F857	JDM		204.6	2
F907	JDM		52.5	<2
F945	JDM		68.7	<2
F945	JDM		44.9	<2
F945	JDM		169.5	2
F945	JDM		146.0	<2
F945	JDM		313.5	2
F946	JDM		6.8	2
F947	JDM		3.7	<3
F948	JDM		20,149.5	100
F948	JDM		6,012.5	50
F970	JDM		1.1	<2
F885	JSLE		21.4	2
F472	JSLE		4,379.4	100

FCL, familial chilblain lupus; SAVI, STING-associated vasculopathy with onset in infancy; SMS, Singleton–Merten syndrome.



Table S8. Information on data used for Fig. 3 (C-E)

Patients	Phenotypes	Gene	IFN $\alpha$	ISG score
			<i>fg/ml</i>	
F759	AGS	<i>ADAR1</i>	111.2	18.6
F788	AGS	<i>ADAR1</i>	36.5	4.6
F755_1	AGS-like	<i>IFIH1</i>	218.5	31.9
F755_2	FCL-like	<i>IFIH1</i>	10,188.8	59.8
F755_3	SMS-like	<i>IFIH1</i>	38.7	16.7
F427	AGS	<i>RNASEH2A</i>	71.4	9.7
F427	AGS	<i>RNASEH2A</i>	47.7	6.5
F1023	AGS	<i>RNASEH2B</i>	2,223.5	6.0
F1169	AGS	<i>RNASEH2B</i>	306.4	7.0
F127	AGS	<i>RNASEH2B</i>	30.4	5.2
F903	AGS	<i>RNASEH2B</i>	248.6	7.4
F190	AGS	<i>RNASEH2B</i>	59.2	5.4
F711	AGS	<i>RNASEH2B</i>	5.3	1.3
F088_1	AGS	<i>RNASEH2C</i>	310.4	8.6
F088_2	AGS	<i>RNASEH2C</i>	506.1	9.8
F559	AGS	<i>SAMHD1</i>	7,699.8	16.2
F1119	AGS	<i>SAMHD1</i>	92.0	17.4
F1081_1	FLC	<i>TREX1</i>	175.4	6.2
F1081_2	Asymptomatic	<i>TREX1</i>	56.4	7.7
F769	AGS	<i>TREX1</i>	3,379.5	16.8
F958	AGS	<i>TREX1</i>	1,221.4	3.6
F831	SAVI	<i>TMEM173</i>	1,657.0	63.7
F831	SAVI	<i>TMEM173</i>	1,989.1	36.3
F831	SAVI	<i>TMEM173</i>	3,847.1	37.5
F831	SAVI	<i>TMEM173</i>	1,395.2	33.7
F831	SAVI	<i>TMEM173</i>	3,316.7	37.6
F831	SAVI	<i>TMEM173</i>	3,134.1	21.4
F1182	SAVI	<i>TMEM173</i>	942.9	24.5
F876	SAVI	<i>TMEM173</i>	14,735.4	24.9
F948	DMJ		20,149.5	37.1
F1105	DMJ		33.0	31.9
F1193	DMJ		50.0	8.5
F1106	DMJ		7,457.6	18.3
F745	DMJ		48.5	8.1
F1162	DMJ		24.2	9.0
F857	DMJ		196.9	25.9
F1022	DMJ		939.4	18.6
F1051	DMJ		14.3	17.8
F918	DMJ		26.7	8.5
F976	DMJ		0.7	0.4
F1175	DMJ		119.8	43.1
F938	DMJ		94.1	9.2
F907	DMJ		1.6	2.4
F940	DMJ		364.5	46.7
F1073	DMJ		92.7	10.2
F1011	DMJ		106.7	17.7
F1093	DMJ		212.6	29.8
F1091	DMJ		82.2	12.0
F947	DMJ		3.7	16.1
F1511	DMJ		116.6	22.0
F1067	DMJ		33.1	4.1
F946	DMJ		19.1	2.7
F945	DMJ		153.4	4.6
F1017	JSLE		11.6	1.9
F1054	JSLE		986.3	24.0
F1078	JSLE		112.3	11.4
F898	JSLE		226.5	11.3
F472	JSLE		4,379.4	25.9
F885	JSLE		21.4	5.4
LUP022-1	SLE		190.4	28.6
LUP023-1	SLE		0.7	0.5
LUP024-1	SLE		0.7	0.7

Table S8. Information on data used for Fig. 3 (C–E) (Continued)

Patients	Phenotypes	Gene	IFN $\alpha$	ISG score
LUP025-1	SLE		1,969.7	27.3
LUP026-1	SLE		51.0	16.1
LUP027-1	SLE		1,204.6	22.8
LUP028-1	SLE		31.7	12.2
LUP029-1	SLE		0.7	25.1
LUP030-1	SLE		0.7	1.9
LUP032-1	SLE		217.5	16.7
LUP034-1	SLE		580.5	24.1
LUP035-1	SLE		0.7	1.1
LUP036-2	SLE		0.7	0.3
LUP037-1	SLE		0.7	0.8
LUP039-2	SLE		0.7	0.2
LUP040-1	SLE		49.0	12.4
LUP042-1	SLE		9.3	6.6
LUP043-1	SLE		15.9	6.8
LUP044-1	SLE		0.7	0.2
LUP045-1	SLE		0.7	0.2
LUP046-1	SLE		83.7	0.3
LUP048-1	SLE		113.6	17.9
LUP049-1	SLE		694.2	19.2
LUP050-1	SLE		0.7	0.3
LUP051-1	SLE		6.0	5.0

FCL, familial chilblain lupus; SAVI, STING-associated vasculopathy with onset in infancy; SMS, Singleton–Merten syndrome.

Table S9. Demographic and clinical associations of raised serum IFN $\alpha$  levels in SLE patients

Parameter	Serum IFN concentration			P-value
	Low ( <i>n</i> = 27)	Medium ( <i>n</i> = 14)	High ( <i>n</i> = 8)	
Age (yr)	47 $\pm$ 2.8	51 $\pm$ 4.2	44 $\pm$ 4.3	0.47
Sex	Female 25, male 2	Female 13, male 1	Female 8, male 0	
CRP	7.8 $\pm$ 2.3	2.7 $\pm$ 0.6	5.9 $\pm$ 2.4	0.20
Anti-dsDNA titer (U/ml)	32.6 $\pm$ 7.4	28.9 $\pm$ 8.5	47.7 $\pm$ 24.9	0.58
Number of IM drugs	1.6	1.6	1.9	0.76

Mean values  $\pm$  SEM are shown with results of one-way ANOVA. dsDNA, double-stranded DNA; IM, immunomodulatory (includes prednisolone, hydroxychloroquine, methotrexate, rituximab, mycophenolate mofetil, and azathioprine).

Table S10. Circulating leukocyte frequencies in STING, AGS, JDM, and controls

Cell subset	Control ( <i>n</i> = 4)	DMJ ( <i>n</i> = 3)	AGS ( <i>n</i> = 4)	STING ( <i>n</i> = 3)	LUPUS ( <i>n</i> = 3)
CD4	12.9 $\pm$ 8.8	9.0 $\pm$ 2.9	24.0 $\pm$ 22.3	17.0 $\pm$ 4.9	34.5 $\pm$ 8.1
CD8	34.6 $\pm$ 17.4	9.3 $\pm$ 3.4	8.9 $\pm$ 4.8	15.8 $\pm$ 5.0	24.5 $\pm$ 11.2
B cells	7.4 $\pm$ 3.1	7.8 $\pm$ 9.1	21.4 $\pm$ 21.3	8.6 $\pm$ 5.4	7.5 $\pm$ 4.4
NK cells	14.9 $\pm$ 4.4	12.0 $\pm$ 9.0	4.9 $\pm$ 4.4	3.9 $\pm$ 4.4	18.1 $\pm$ 14.5
Mo	33.2 $\pm$ 13.2	48.7 $\pm$ 17.9	17.5 $\pm$ 14.6	3.4 $\pm$ 1.7	4.2 $\pm$ 5.5
pDCs	0.7 $\pm$ 0.5	1.4 $\pm$ 0.7	0.9 $\pm$ 1.1	0.3 $\pm$ 0.2	0.1 $\pm$ 0.0

Values are reported as mean  $\pm$  SD. Mo, monocytes.

Table S11. Sorted cell numbers and IFN concentration

Diseases and patients	Subsets	Cells (50 µl lysate)	fg/ml	attograms/cell
<b>JDM</b>				
F1073	T4	30,000	4.16	0.0069
	T8	24,000	2.20	0.0046
	NK	20,000	1.68	0.0042
	B	NA	NA	NA
	Mo	30,000	<LOD	ND
	pDC	11,000	<LOD	ND
F948	T4	60,000	8.88	0.0074
	T8	90,500	17.02	0.0094
	NK	60,000	3.53	0.0029
	B	90,000	7.05	0.0039
	Mo	79,000	<LOD	ND
	pDC	4,000	3.67	0.0458
F857	T4	52,000	10.87	0.0104
	T8	45,000	20.83	0.0231
	NK	23,000	5.41	0.0118
	B	23,000	5.85	0.0127
	Mo	100,000	2.38	0.0012
	pDC	3,000	<LOD	ND
<b>AGS</b>				
F1119	T4	30,000	3.14	0.0052
	T8	16,500	<LOD	ND
	NK	4,000	<LOD	ND
	B	6,000	<LOD	ND
	Mo	30,000	<LOD	ND
	pDC	3,800	<LOD	ND
F1023	T4	60,000	21.81	0.0182
	T8	34,000	28.58	0.0420
	NK	5,000	2.37	0.0237
	B	60,000	19.12	0.0159
	Mo	66,000	4.66	0.0035
	pDC	1,700	<LOD	ND
F88-1	T4	60,000	27.70	0.0231
	T8	100,000	32.48	0.0162
	NK	210,000	66.61	0.0159
	B	85,000	10.94	0.0064
	Mo	250,000	15.85	0.0032
	pDC	9,000	<LOD	ND
F559	T4	300,000	43.90	0.0073
	T8	200,000	56.40	0.0141
	NK	210,000	49.08	0.0117
	B	170,000	25.51	0.0075
	Mo	370,000	16.60	0.0022
	pDC	5,300	<LOD	ND
<b>SAVI</b>				
F876	T4	20,000	8.82	0.0220
	T8	5,000	11.65	0.1165
	NK	20,000	15.09	0.0377
	B	10,200	2.56	0.0126
	Mo	25,000	1,126.72	2.2534
	pDC	35,000	1,856.51	2.6522
F1429	T4	100,000	15.20	0.0076
	T8	100,000	26.36	0.0132
	NK	30,000	13.50	0.0225
	B	30,000	5.31	0.0088
	Mo	90,000	1,337.37	0.7430
	pDC	6,000	219.22	1.8268
F1802	T4	100,000	18.15	0.0091
	T8	93,000	13.43	0.0072
	NK	100,000	9.70	0.0049
	B	100,000	19.18	0.0096
	Mo	100,000	198.49	0.0992
	pDC	16,000	36.75	0.1148

Table S11. Sorted cell numbers and IFN concentration (Continued)

Diseases and patients	Subsets	Cells (50 µl lysate)	fg/ml	attograms/cell
<b>SLE</b>				
LU3	T4	100,000	22.74	0.0114
	T8	100,000	2.61	0.0013
	NK	100,000	5.61	0.0028
	B	111,833	43.81	0.0196
	Mo	3,452	<LOD	ND
	pDC	1,317	<LOD	ND
LU4	T4	100,000	7.35	0.0037
	T8	46,839	7.08	0.0076
	NK	56,442	26.51	0.0235
	B	4,746	<LOD	ND
	Mo	33,909	1.17	0.0017
	pDC	588	<LOD	ND
LU6	T4	100,000	76.47	0.0382
	T8	100,000	<LOD	0.0010
	NK	100,000	2.38	0.0012
	B	100,000	8.36	0.0042
	Mo	21,020	<LOD	ND
	mDC	1,614	<LOD	ND
<b>Controls</b>				
CT1	T4	100,000	15.62	0.0078
	T8	50,000	14.08	0.0141
	NK	50,000	3.26	0.0033
	B	50,000	11.53	0.0115
	Mo	510,000	5.74	0.0006
	pDC	9,000	4.01	0.0223
CT2	T4	100,000	24.06	0.0120
	T8	100,500	28.80	0.0143
	NK	100,000	23.49	0.0117
	B	100,000	29.28	0.0146
	Mo	100,000	1.47	0.0007
	pDC	16,000	15.54	0.0486
CT3	T4	100,000	24.84	0.0124
	T8	59,500	17.44	0.0147
	NK	66,500	5.13	0.0039
	B	51,000	37.89	0.0371
	Mo	54,000	5.53	0.0051
	pDC	15,000	4.39	0.0146

B, B cell; Mo, monocyte; mDC, myeloid DC.

Table S12. Clinical information of patients studied for cell subset IFNα content

Patient	Phenotype	Disease activity	Treatment	Clinical response to treatment	IFNα serum
F857	JDM	High	Steroids/HCO	Poor	fg/ml 198
F948	JDM	High	MMF/HCO	Poor	3,552
F1073	JDM	High		Poor	32
F876	SAVI	Low	Ruxolitinib	Good	43,031
F1429	SAVI	Moderate	Ruxolitinib	Good	896
F1802	SAVI	Low	Not treated		44,579
F1023	AGS	Severe developmental delay	Not treated	Not relevant	581
F1119	AGS	Moderate developmental delay with active chilblains	Not treated	Not relevant	149
F559	AGS	Moderate developmental delay with active chilblains	Not treated	Not relevant	622
F88	AGS	Severe developmental delay	Not treated	Not relevant	739
LU3	SLE	Joint/renal involvement	Prednisolone	Good	241
LU4	SLE	Joint/renal/lung involvement	Prednisolone/HQ/MMF	Good	793
LU6	SLE	Joint/renal involvement	Prednisolone/MMF	Good	131

HCO, hydroxychloroquine; MMF, mycophenolate mofetil; SAVI, STING-associated vasculopathy with onset in infancy.