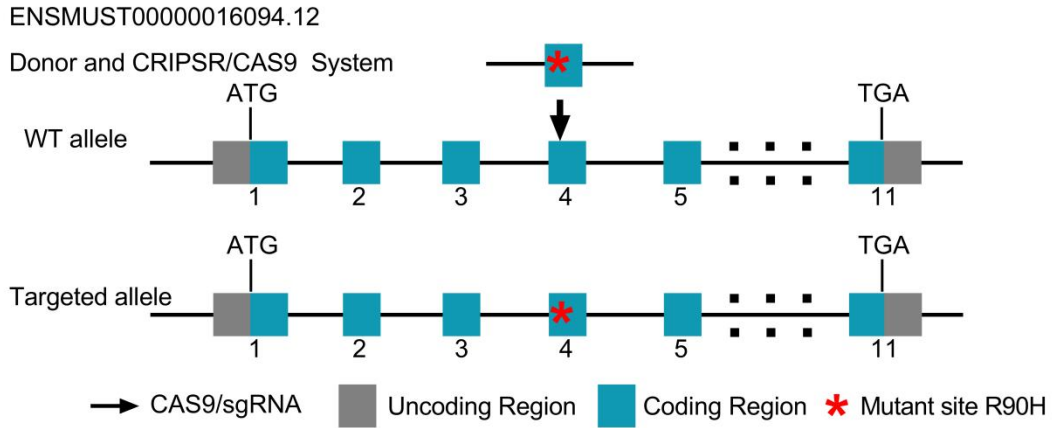
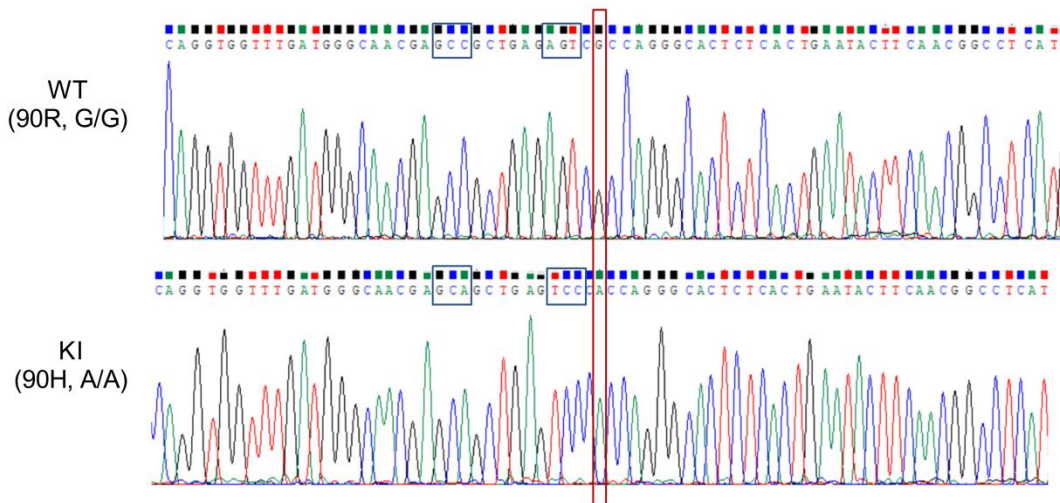


1 **Supplemental Figures**

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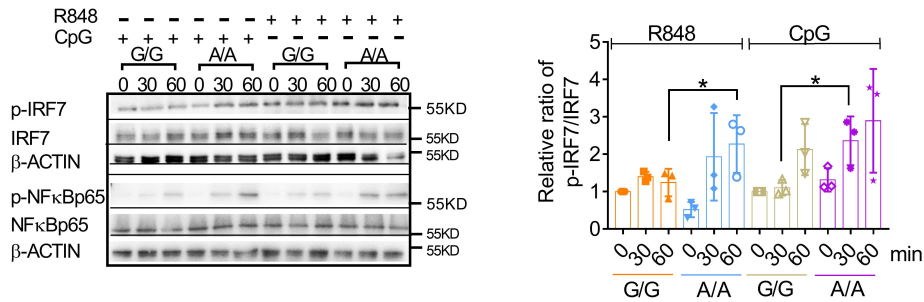
3 **Supplemental Figure 1. Strategy of constructing KI mice.** (A) KI mice were
4 generated by using CRISPR/CAS9 gene editing and homologous recombination
5 technique. (B) Verification of p.90R to p.90H mutation in KI mice. The black squares
6 indicate the position of synonymous codon substitution.

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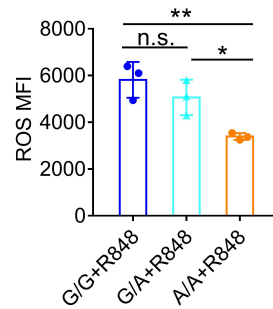
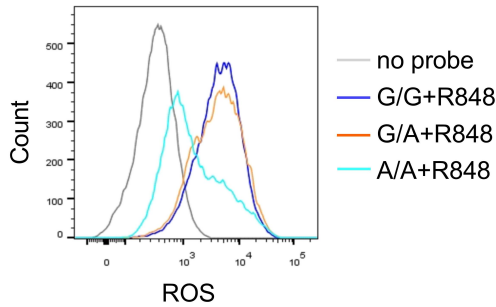
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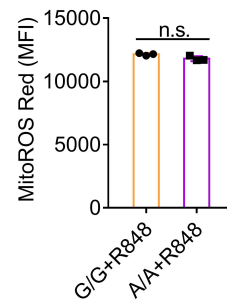
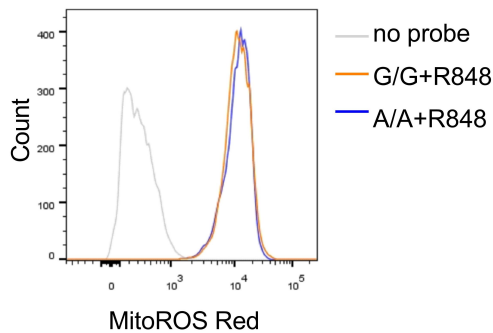
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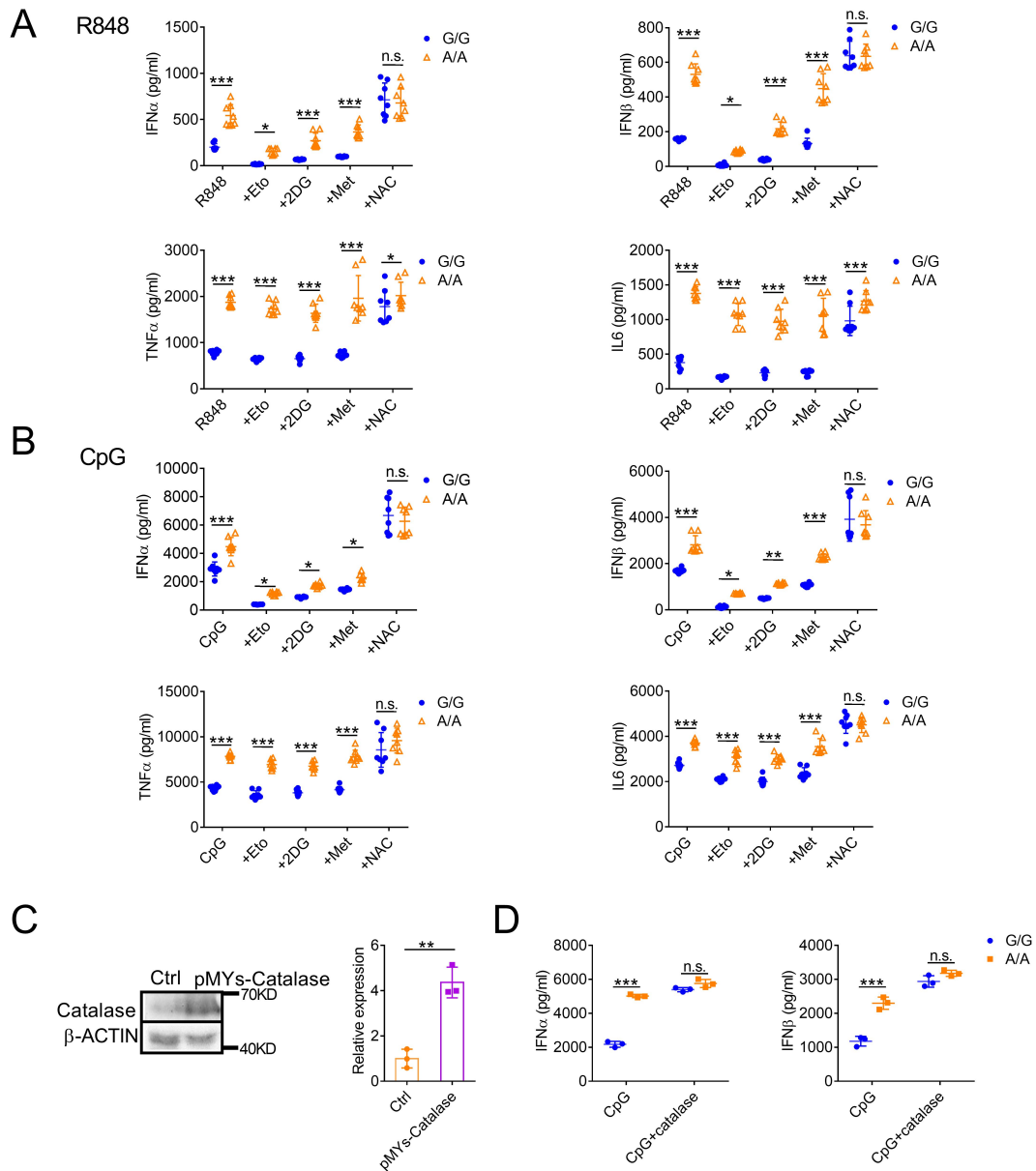


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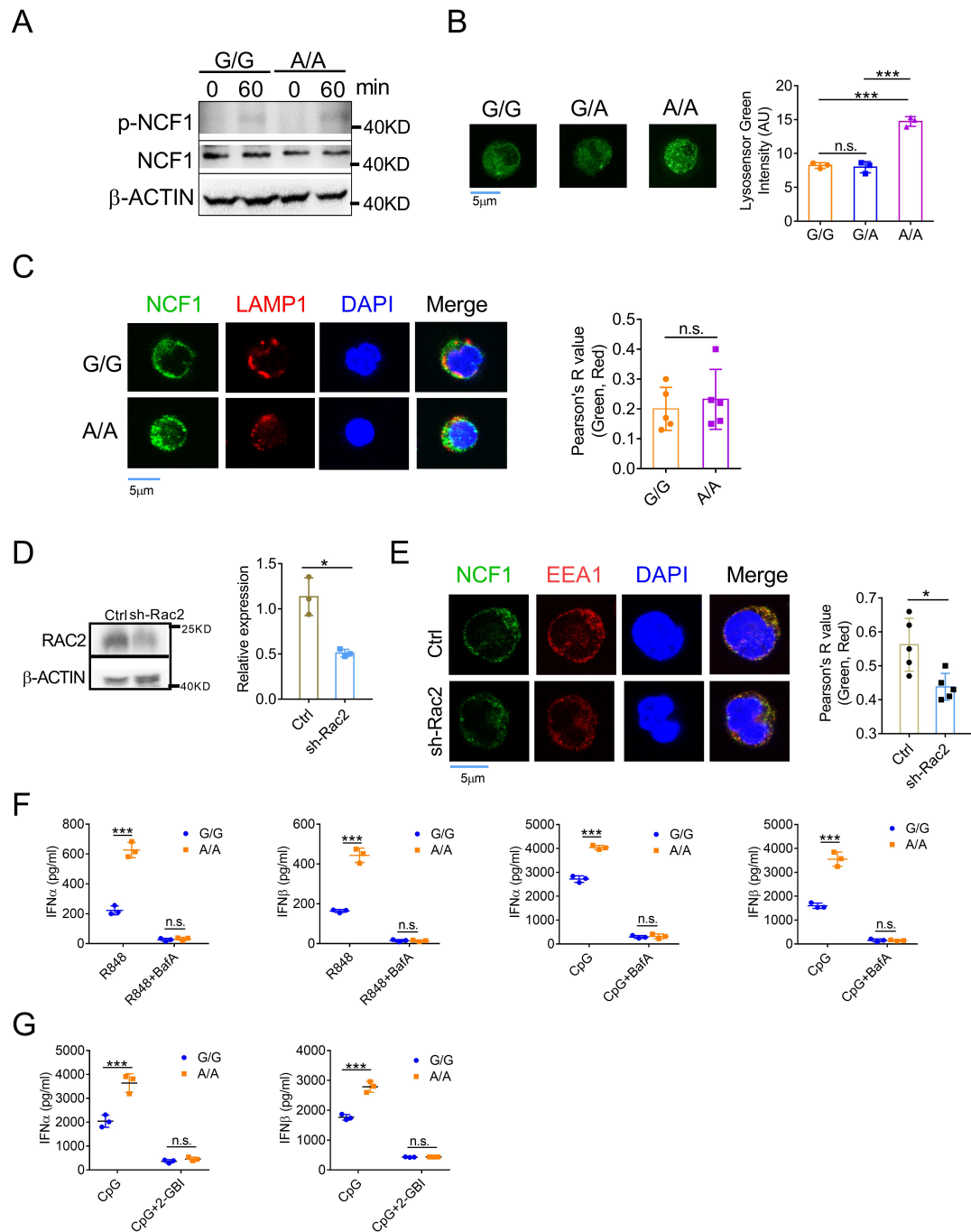
11 **Supplemental Figure 2. The activation and ROS levels of pDCs with different**
 12 **alleles.** (A) G/G and A/A pDCs were generated by FLT3L induction. Then, pDCs
 13 were sorted and stimulated with R848 or CpG for indicated time. The levels of
 14 phospho-IRF7 (p-IRF7), IRF7, phospho-NFκB p65 (p-NFκBp65) and NFκBp65
 15 were detected. β-ACTIN was used as loading control. *p* value was determined by a
 16 two-tailed paired *t* test. Error bars represent SEM. (**p*<0.05). *n*=3. (B, C) Splenic
 17 pDCs from G/G, G/A and A/A mice were stimulated with R848 for 1 hour and then
 18 analyzed by FACS. The levels of (B) ROS and (C) mitochondrial ROS are shown. *p*
 19 values were determined by one-way ANOVA with Tukey's multiple comparison test.
 20 Error bars represent SEM. (**p*<0.05, ***p*<0.01, n.s. not significant).



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22 **Supplemental Figure 3. ROS scavenger eliminates the difference in the**
 23 **production of IFN-I resulted from NCF1 p.R90H.** PDCs were pretreated with or
 24 without etomoxir (Eto), 2-DG, metformin (Met), or NAC for 2 hours and then
 25 stimulated with (A) R848 or (B) CpG for 24 hours. The expression of IFN α , IFN β ,
 26 TNF α and IL6 were detected. n=8. (C, D) Bone marrow progenitors from WT or KI
 27 mice were transfected with empty or Catalase overexpressing lentivirus, and followed
 28 by 8-day FLT3L induction. PDCs were sorted and stimulated with CpG for 24 hours.
 29 (C) The overexpression of Catalase. *p* value was determined by a two-tailed paired *t*
 30 test. Error bars represent SEM. (**p*<0.05). n=3. (D) The expression of IFN α and IFN β .
 31 n=3. *p* values were determined by two-way ANOVA. Error bars represent SEM.
 32 (**p*<0.05, ***p*<0.01, n.s. not significant).

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 35 **Supplemental Figure 4. The mechanism of NCF1 p.R90H variation on the**
 36 **activation of pDCs.** (A-C) G/G, G/A and A/A pDCs generated by FLT3L induction in
 37 vitro were stimulated with CpG for 1 hour. (A) The levels of phospho-NCF1 (p-NCF1)
 38 and NCF1 were detected. β-ACTIN was used as loading control. (B) The pH of late
 39 endosome/lysosome was indicated by lysosome sensor, detected by con-focal, and
 40 then measured by Image J. One point indicates mean of Pearson's R values from 10
 41 cells and total of 30 cells per group were calculated. Scale bar represents 5 μm. p
 42 values were determined by one-way ANOVA with Tukey's multiple comparison test.
 43 (C) The co-localization of NCF1 (green) and LAMP1 (Red) was detected by confocal
 44 microscopy and analyzed by Image J. (D, E) Lineage^{c-kit}^{hi} bone marrow progenitors

45 were isolated from WT mice by using FACS. Lentivirus expressing shRNA specific
46 targeting *Rac2* was transduced into bone marrow progenitors. Cells were cultured
47 with 200ng/mL FLT3L for 8 days. Then pDCs were sorted and subjected to further
48 study. (D) Western blot detection of RAC2. n=3. *p* values were determined by a
49 two-tailed paired *t* test. (E) PDCs transfected with empty (Ctrl) or shRNA lentivirus
50 targeting *Rac2* (sh-*Rac2*) were stimulated with CpG for 2 hours. The co-localization
51 of NCF1 (green) and EEA1 (Red) was detected by confocal microscopy and analyzed
52 by Image J. One point indicates mean of Pearson's R values from 6 cells and total of
53 30 cells per group were calculated. Scale bar represents 5 μ m. *p* values were
54 determined by a one or two-tailed unpaired *t* test. Error bars represent SEM. (n.s. not
55 significant). (F) PDCs were pre-treated with or without 100nM bafilomycin A (BafA)
56 for 1 hour and then stimulated with 10 μ g/mL R848 or 0.5 μ M ODN2216 (CpG) for 24
57 hours. (G) PDCs were pre-treated with or without 2-GBI for 1 hour and then
58 stimulated with 0.5 μ M ODN2216 (CpG) for 24 hours. The expression of type I IFNs
59 in supernatant was detected by ELISA. n=3. Data were analyzed by using two-way
60 ANOVA. Error bars represent SEM. (**p*<0.05, ****p*<0.001, n.s. not significant).

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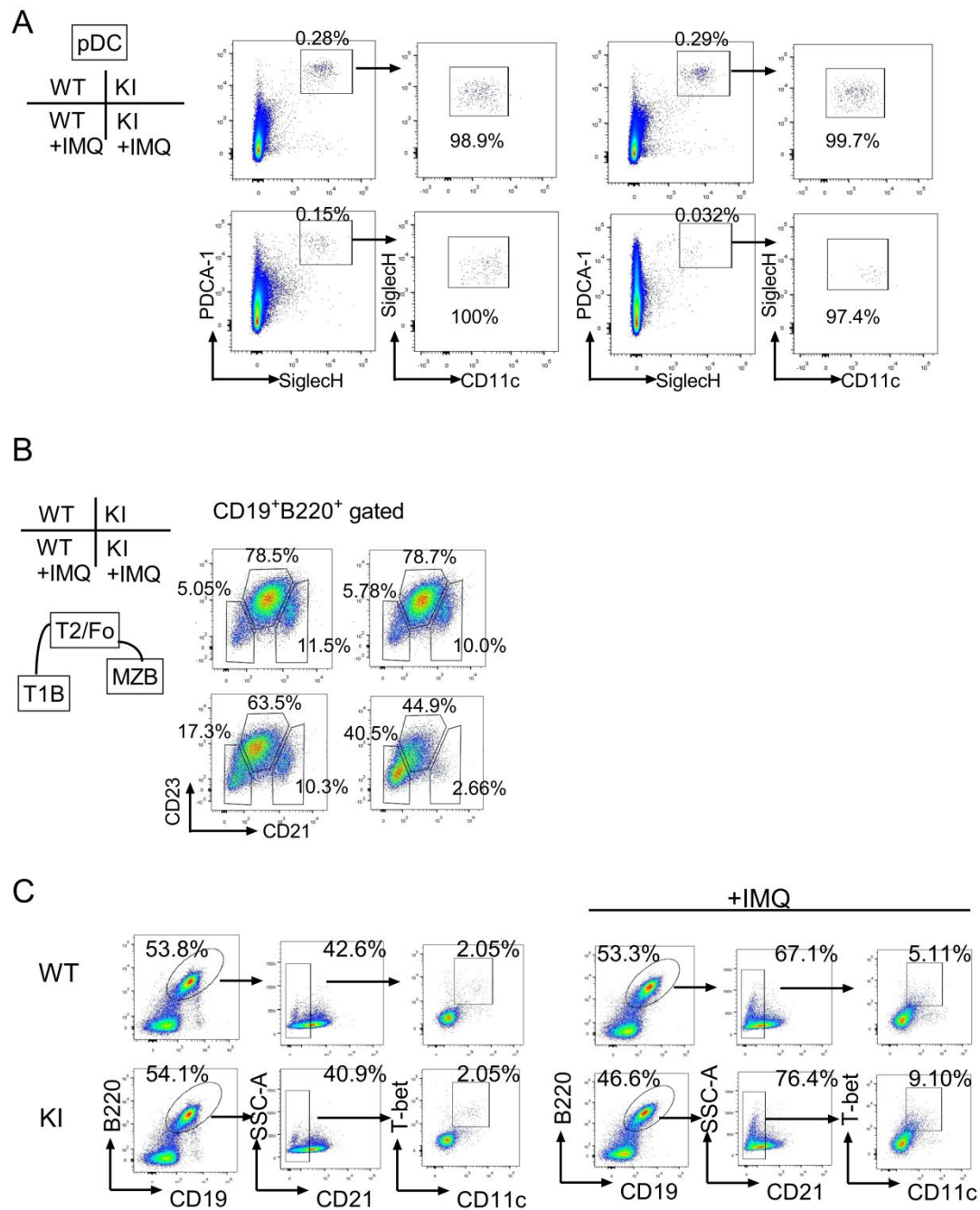
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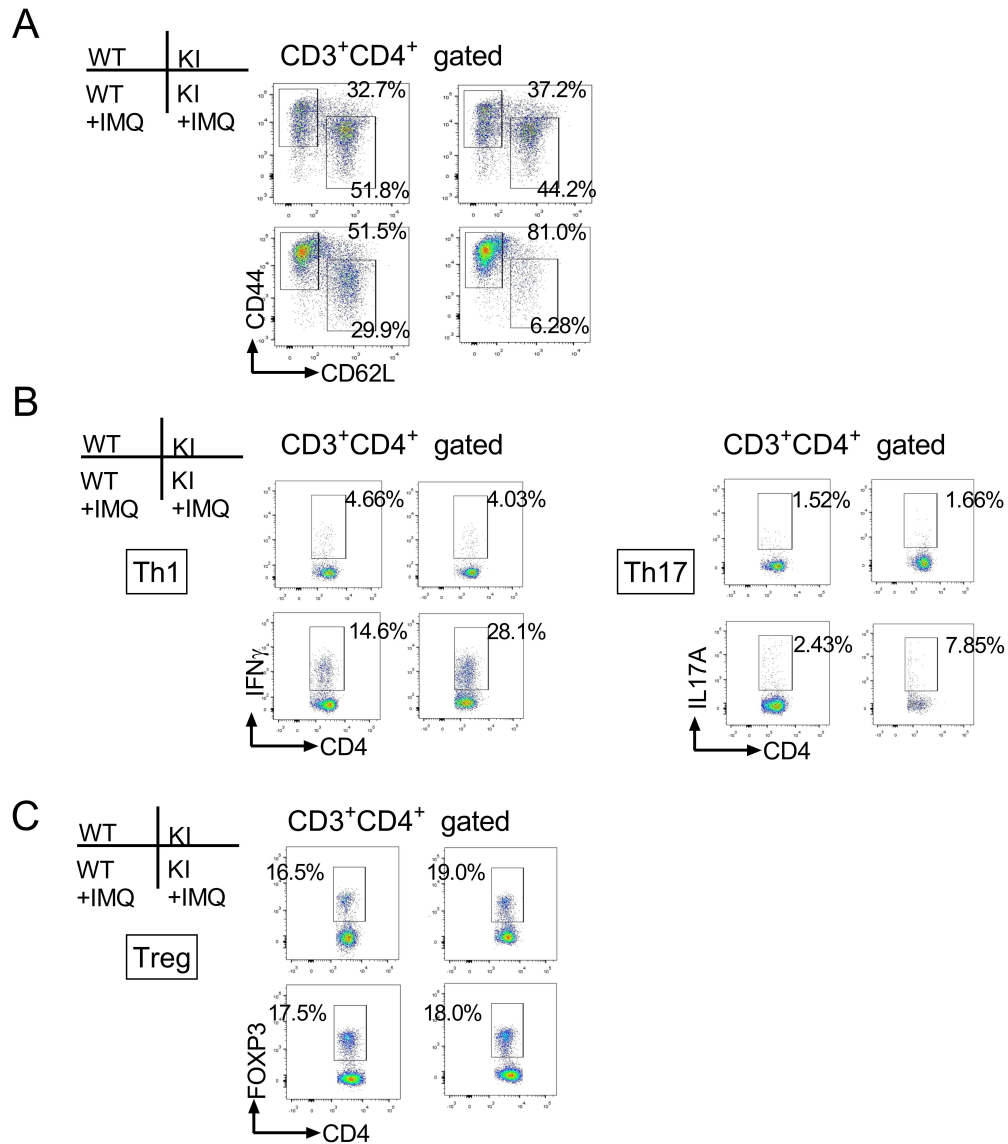
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76 **Supplemental Figure 5. NCF1 p.R90H facilitates the aberrant activation of pDC**
 77 **and B cells.** The cellular gating strategies of (A) pDCs (CD11c^{int}, SiglechH⁺, PDCA-1⁺)
 78 and (B) B cells subsets, including MZB (CD19⁺, B220⁺, CD21^{hi}, CD23⁻), T2/Fo B
 79 cells (CD19⁺, B220⁺, CD21^{lo}, CD23^{hi}), T1 B cells (CD19⁺, B220⁺, CD21⁻, CD23⁻) and
 80 (C) ABC cells (CD19⁺, B220⁺, CD21⁻, CD11c⁺, T-bet⁺) in Figure 4 are shown.



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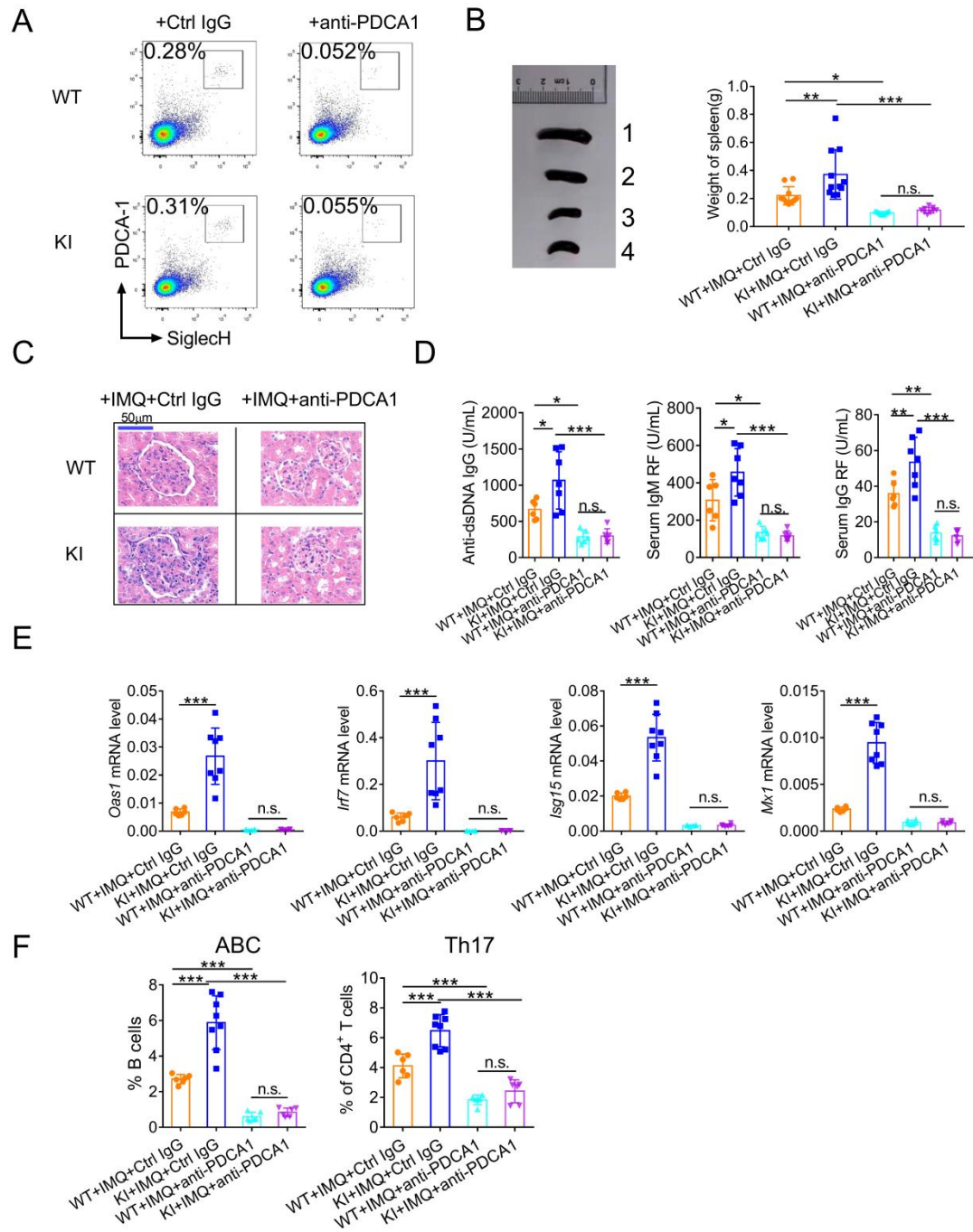
82 **Supplemental Figure 6. NCF1 p.R90H facilitates the aberrant activation of T**

83 **cells.** (A) The FACS analysis of naïve T cells (CD3⁺, CD4⁺, CD44^{lo}, CD62L^{hi}),

84 activated T cells (CD3⁺, CD4⁺, CD44^{hi}, CD62L^{lo}), (B) Th1 (CD3⁺, CD4⁺, IFN γ ⁺),

85 Th17 (CD3⁺, CD4⁺, IL-17A⁺) and (C) Treg (CD3⁺, CD4⁺, Foxp3⁺) in Figure 4 are

86 shown.

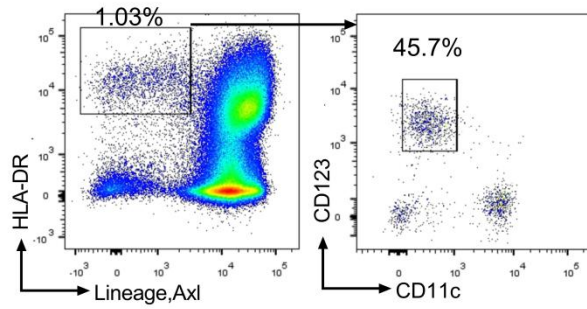


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 88 **Supplemental Figure 7. PDC depletion eliminates the pathogenic aggravation**
 89 **caused by NCF1 p.R90H.** (A) WT and KI mice were intraperitoneally pre-treated
 90 with anti-PDCA1 antibody (anti-PDCA1) or rat IgG2b isotype control (Ctrl IgG) 4
 91 days before IMQ application. The efficiency of pDC depletion. (B-F) Mice were then
 92 treated with IMQ, together with Ctrl IgG or anti-PDCA1 antibodies for 8 weeks. (B)
 93 The picture and the weight of spleen. n=9-11. 1, WT+IMQ+Ctrl IgG; 2,
 94 KI+IMQ+Ctrl IgG; 3, WT+IMQ+anti-PDCA1; 4, KI+IMQ+anti-PDCA1. (C) H&E
 95 staining of kidney. (D) The levels of anti-dsDNA antibodies, serum IgM RF and IgG
 96 RF. (E) The expression of *Oas1*, *Irf7*, *Isg15* and *Mx1* in the kidney of mice. (F) FACS
 97 analysis of ABCs and Th17 cells in the spleen. n=6-8. Data were analyzed by using

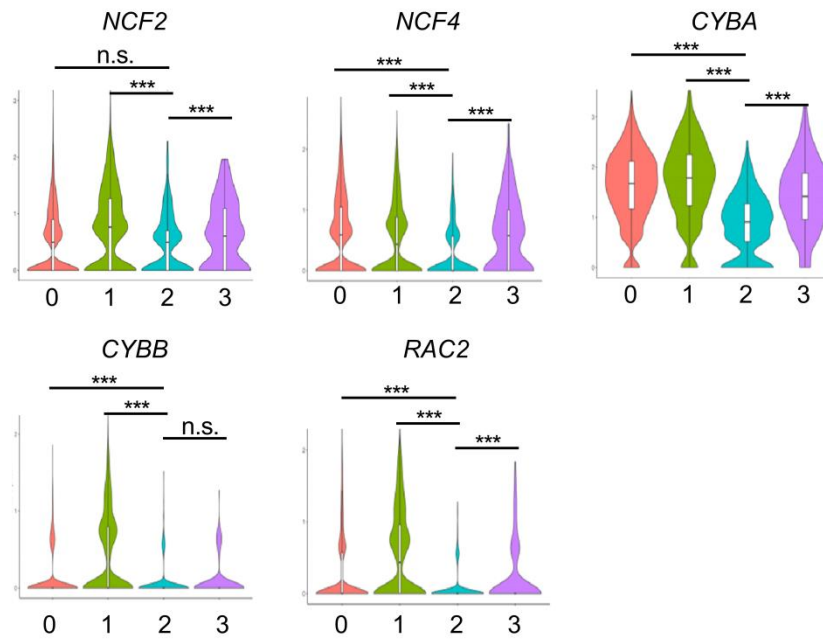
98 one-way ANOVA with Tukey's multiple comparison test. Error bars represent SEM.
99 (*p<0.05, **p<0.01, ***p<0.001, n.s. not significant).

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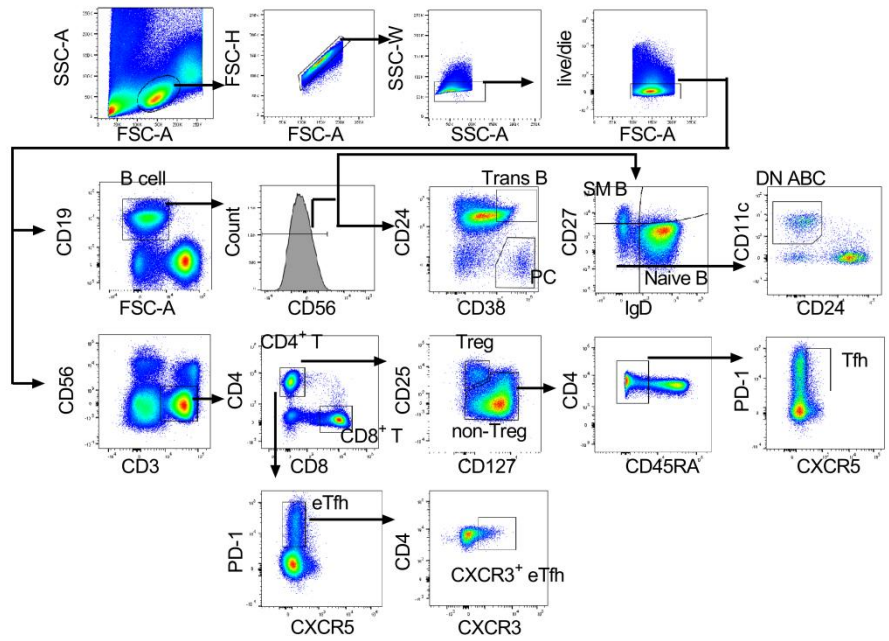
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Supplemental Figure 8. Sorting strategy of human primary pDCs. (A) pDCs were sorted as Lineage⁻ (CD3/CD14/CD16/CD19/CD20/CD56), HLA-DR⁺, CD11c⁻, AXL⁻, CD123⁺. The purity of pDCs was >99.9%. (B) The expression of *NCF2*, *NCF4*, *CYBA*, *CYBB* and *RAC2* in pDC subpopulations. *p* values were determined by using Wilcoxon test. (***)*p*<0.001).



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149 **Supplemental Figure 9. Gating strategies of immune cells subsets from SLE**
 150 **patients.** Samples in Figure 6 were analyzed by FACS using indicated markers.

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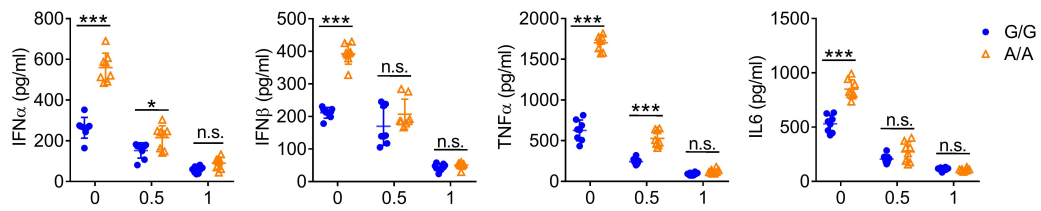
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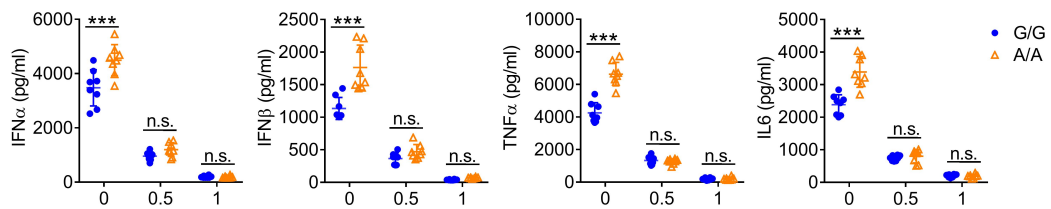
A

R848



B

CpG



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Supplemental Figure 10. HCQ alleviates the excessive activation of pDCs

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resulted from NCF1 p.R90H. (A, B) Splenic pDCs were pre-treated with 0, 0.5 or 1

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μ M HCQ for 2 hours, and then stimulated with R848 or CpG for 24 hours. The

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expression of IFN α , IFN β , TNF α and IL6 was detected. n=8. *p* values were

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determined by two-way ANOVA. Error bars represent SEM. (p*<0.05, ***p*<0.01,**

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******p*<0.001, n.s. not significant).**

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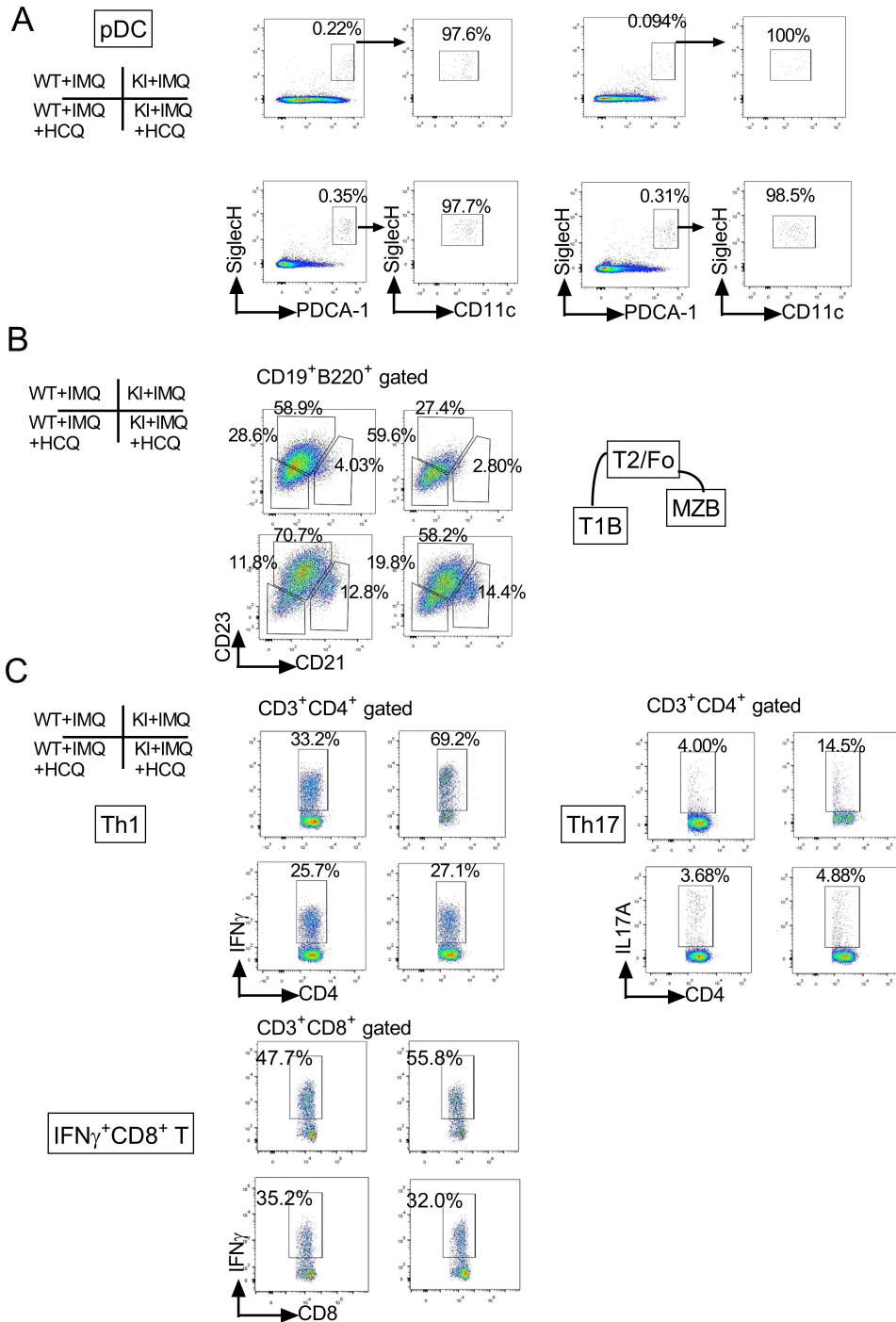
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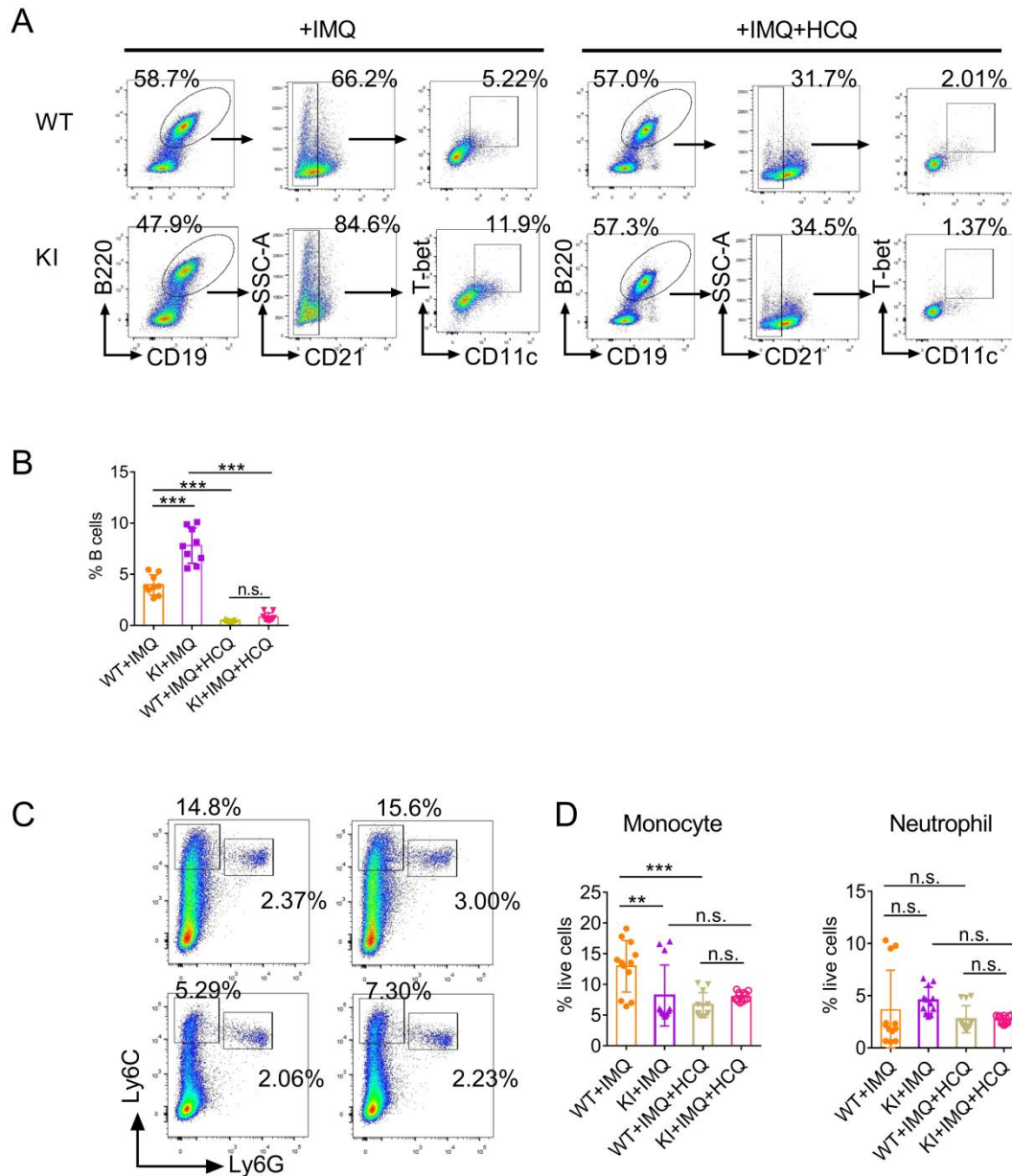


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178 **Supplemental Figure 11. FACS analysis of immune cells suppressed by HCQ.** The
 179 FACS gating strategies of (A) pDCs (CD11c^{int}, Siglech⁺, PDCA-1⁺), (B) B cell
 180 subsets, including MZB (CD19⁺, B220⁺, CD21^{hi}, CD23⁻), T1 B (CD19⁺, B220⁺,
 181 CD21⁻, CD23⁻) and T2/Fo B cells (CD19⁺, B220⁺, CD21^{lo}, CD23^{hi}), and (C) T cell
 182 subsets, including Th1 (CD3⁺, CD4⁺, IFN γ ⁺), Th17 (CD3⁺, CD4⁺, IL17A⁺) and IFN γ ⁺
 183 CD8⁺ T cells (CD3⁺, CD8⁺, IFN γ ⁺) in Figure 8 are shown.

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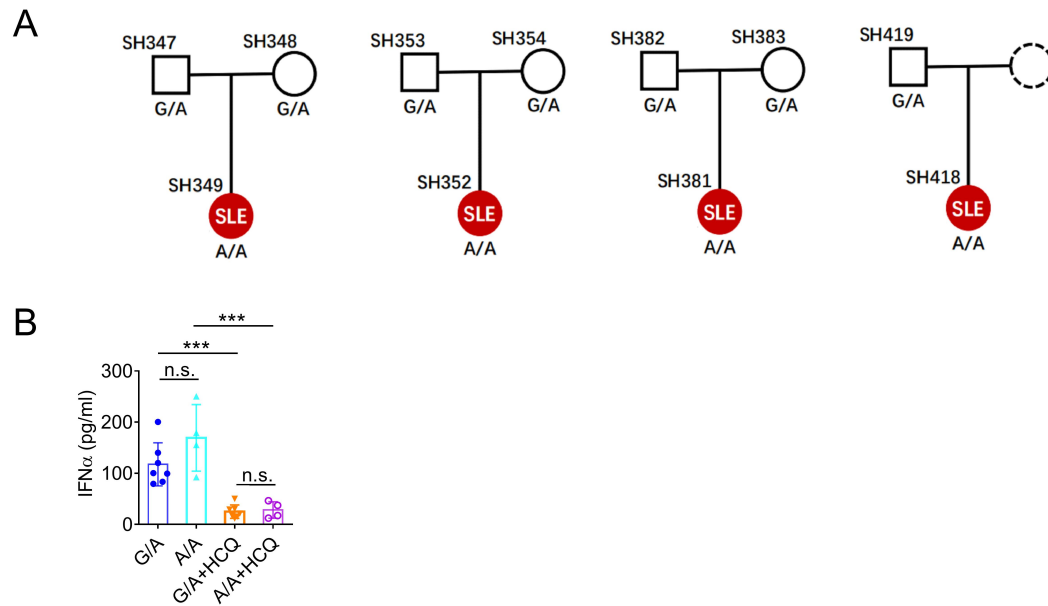


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187 **Supplemental Figure 12. Impact of NCF1 p.R90H and HCQ on ABCs, monocytes**
 188 **and neutrophils.** WT and KI mice were treated with IMQ, together with or without
 189 HCQ for 8 weeks. Splenocytes were isolated and analyzed by FACS. (A) Gating
 190 strategy of ABCs (CD19⁺, B220⁺, CD21⁻, CD11c⁺, T-bet⁺). n=9. (B) The proportion of
 191 ABCs. (C) FACS analysis of monocytes (Ly6C^{hi}, Ly6G⁻) and neutrophils (Ly6C^{lo},
 192 Ly6G⁺). (D) Statistics of monocytes and neutrophils. *p* values were determined by
 193 one-way ANOVA with Tukey's multiple comparison test. Error bars represent SEM.
 194 (***p*<0.01, ****p*<0.001, n.s. not significant).

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198 **Supplemental Figure 13. The therapeutic effect of HCQ on PBMCs from SLE**

199 **patients with NCF1 p.R90H.** (A) The genetic map of SLE families. (B) PBMCs

200 were pre-incubated with 0.5 μ M HCQ for 2 hours and stimulated with CpG for 24

201 hours. The expression of IFN α was detected. SH indicates sample number. *p* values

202 were determined by one-way ANOVA with Tukey's multiple comparison test. Error

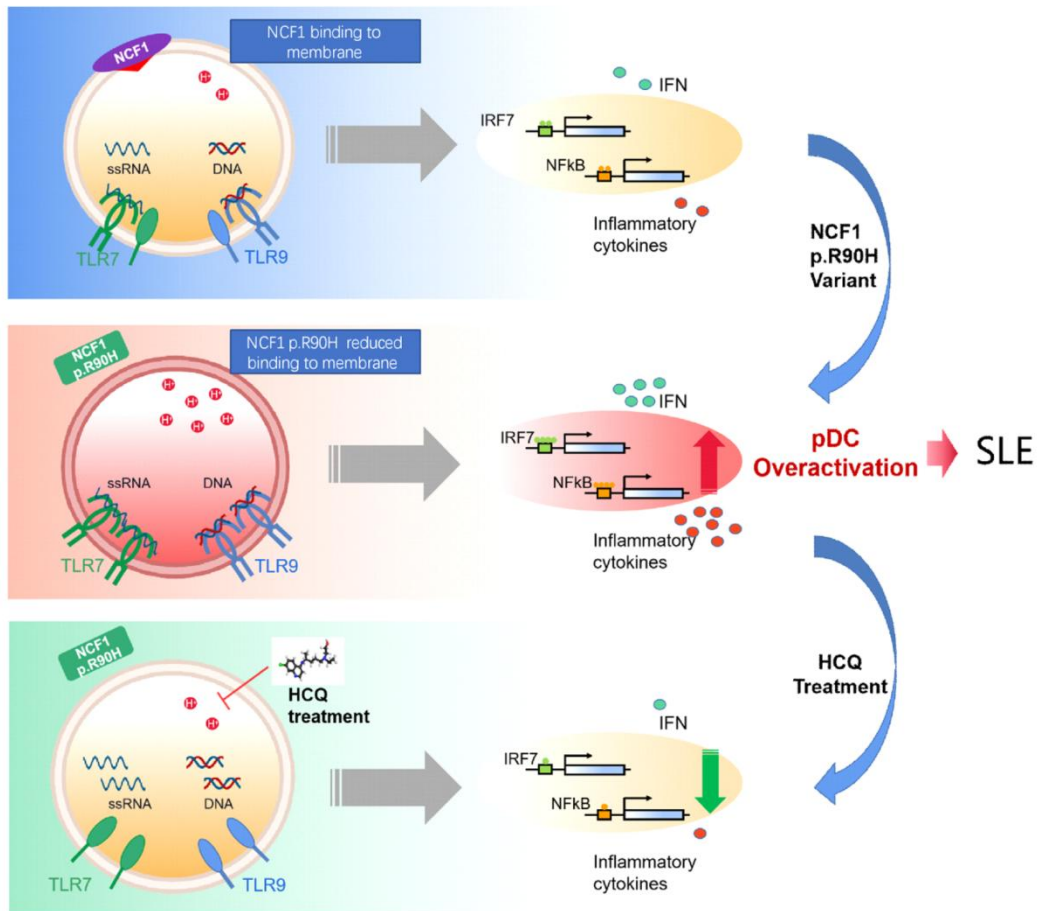
203 bars represent SEM. (***)*p*<0.001, n.s. not significant).

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209 **Supplemental Figure 14. Regulation model.** NCF1 p.R90H variant leads to
 210 impaired localization of NCF1 on the endosomal membrane, followed by acidified pH
 211 and more cleaved activation of TLR receptors, which facilitates pDC activation and
 212 lupus progression. HCQ application alleviates the aggravation of NCF1 p.R90H on
 213 lupus, and this SNP could serve as a genetic biomarker for HCQ treatment.

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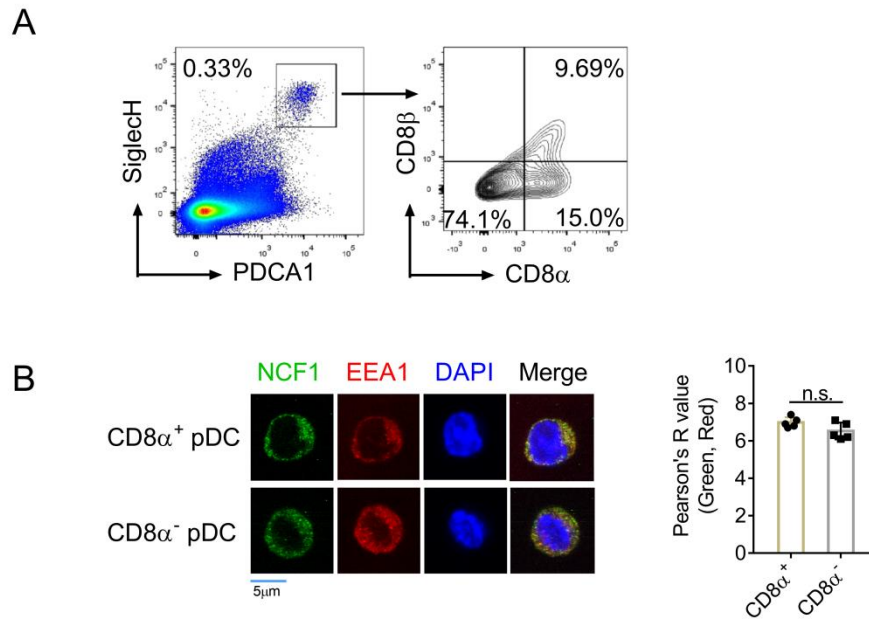
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221 **Supplemental Figure 15. CD8 α^- and CD8 α^+ pDCs show similar endosomal**
 222 **localization of NCF1.** Splenic pDCs subsets were isolated and stimulated with
 223 10 μ g/mL R848 for 1 hour. The colocalization of NCF1 and EEA1 was detected by
 224 fluorescence microscope. (A) The gating strategy of pDC subsets. (B) The
 225 co-localization of NCF1 (green) and EEA1 (Red) was detected by confocal
 226 microscopy and analyzed by Image J. One point indicates mean of Pearson's R values
 227 from 6 cells and total of 30 cells per group were calculated. Scale bar represents 5 μ m.
 228 *p* value was determined by a two-tailed unpaired *t* test. Error bars represent SEM. (n.s.
 229 not significant).

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240 **Supplemental Tables**241 **Supplemental Table 1. Demographic and clinical characteristics of the patients.**

	SLE patients
Number	49
Age (years), mean \pm SD	35.4 \pm 10.5
Female, n (%)	49 (100%)
Disease Duration (months), median (IQR) *	53.8 (105)
System involvement, n (%) * ¶	
Mucocutaneous	12 (30.0%)
Musculoskeletal	3 (7.5%)
Neuropsychiatric	1 (2.5%)
Cardiorespiratory	4 (10.0%)
Renal	19 (47.5%)
Hematological	16 (40.0%)
Disease assessment, median (IQR) *	
SLEDAI	7.0 (6.0)
Laboratory tests	
High ESR, (positive/tested)	24/38
High CRP, (positive/tested)	10/20
Low C3, (positive/tested)	30/40
ANA, (positive/tested)	37/38

Anti-Smith antibody, (positive/tested)	4/38
Anti-RNP antibody, (positive/tested)	14/38
Anti-dsDNA (IU/mL), median (IQR) *	26.2 (41.0)
Treatment, n (%) #	
Prednisone	37 (100%)
Hydroxychloroquine	22 (59.5%)
Immunosuppressants	14 (37.8%)

242 *SLEDAI* Systemic Lupus Erythematosus Disease Activity Index, *ESR* Erythrocyte sedimentation
243 rate, *CRP* C-Reactive Protein, *C3* complement 3, *ANA* anti-nuclear antibody, *RNP*
244 ribonucleoprotein.

245 * Among the 49 patients, 9 patients' clinical information were missing. The percentage calculation
246 was based on 40 patients. # 12 patients' treatment information were missing. The percentage
247 calculation was based on 37 patients. ¶ System involvement indicated all the system involvements
248 during the disease course of the patients.

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Supplemental Table 2. SLE families information.

		23			24			35			47	
sample ID		SH349	SH350	SH351	SH352	SH353	SH354	SH381	SH382	SH383	SH418	SH419
gender		Female	Male	Female	Female	Male	Female	Female	Male	Female	Female	Male
patient_age		6	37	34	11			14	42	37	13	35
relationship		Patient	Father of SH349	Mother of SH349	Patient	Father of SH352	Mother of SH352	Patient	Father of SH381	Mother of SH381	Patient	Father of SH418
Autoimmune disease	SLE	y	n	n	y	n	n	y	n	n	y	n
	Hashimoto's Thyroiditis	n			n			n			n	
	Grave's disease	n			n			n			n	
	T1D	n			n			n			n	
	Vitiligo	n			n			n			n	
	Alopecia	n			n			n			n	
	RA	n			n			n			n	
	Sjogren's Sd	n			n			n			n	
	Vasculitis	n			n			n			n	
Other												
Lupus ACR	Acute Cutaneous lupus	y			y			y			y	

criteria	Chronic cutaneous lupus												
	Aphthous ulcers	n			y			y			y		
	Alopecia	n			n			n			n		
	Arthritis (2 or more joints)	y			y			y			n		
	Serositis	n			n			n			n		
	Renal: Red cell casts	n			y			n			y		
	Renal: >500g protein/24h	n			n			n			y		
	Neurological: Seizures	n			n			n			n		
	Neurological: Psychosis	n			n			n			n		
	Neurological: myelitis	n			n			n			n		
	Autoimmune hemolytic anemia	n			n			n			n		
	Leukopenia:<4.0 ×10 ⁹ /L	n			n			n			n		
	Lymphopenia:<1.0 ×10 ⁹ /L	n			n			y			n		
	Thrombocytopenia:<1.0 ×10 ⁹ /L	n			n			n			n		
	ANA	1: 640			1:160			1: 320					
	dsDNA antibodies (IU/ml)	8.4 IU/M L			28.94 IU/M L			65 IU/ ML			45.98 IU/M L		
	Sm	n			n			y					

	Lupus anticoagulant	n			n			y				
	Cardiolipin antibodies (medium or high titre)	n			n			n				
	beta-2 glycoprotein I antibodies	n			n			n				
	Direct Coombs Test	n			n			n				
	Low C3	n			y			y			y	
	Low C4	n			n			y			y	
Additional lupus phenotypes: a	Raynaud's phenomenon	n			n			n			n	
	Digital vasculitis	n			n			n			n	
	Cutaneous vasculitis	n			n			n			n	
	Arterial thrombosis	n			n			n			n	
	Deep venous thrombosis	n			n			n			n	
	Acute pulmonary embolus	n			n			n			n	
Additional lupus phenotypes: b	Renal	n			n			n			y	
	WHO Glomerulonephritis histological class											
	Nephrotic syndrome	n			n			n			n	
	Highest serum creatinine (umol/L)	24 umol/ L			58 umol/ L			30 umol /L			34 umol/ L	
	Lowest GFR											

Additional	Dry mouth	y			y			y			y	
lupus												
phenotypes: c												
Sicca	Dry eyes											
symptoms		n			n			n			n	
Additional	Tenosynovitis	n			n			n			n	
lupus	Joint subluxation	n			n			y			n	
phenotypes: d												
Musculoskeletal	Myositis											
tal		n			n			n			n	
Additional	Hepatitis	n			n			n			n	
lupus	Mesenteric ischaemia	n			n			n			n	
phenotypes: e												
Gastrointestinal	Pancreatitis											
al		n			n			n			n	
Additional	RNP	n			y			y			n	
lupus	SSA	n			n			y			n	
phenotypes: g	SSB	n			n			y			n	
Serology	Ribosomal P	n			n			n			n	
	Smooth muscle	n			n			n				
	Thyroidperoxidase	n			n			n				

	Thyroglobulin	n			n			n				
	Gastric parietal cell	n			n			n				
	Skin	n			n			y			y	
	Hypergammaglobulinaemia	n			n			n				
	Other											

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262 “y” = Yes, “n” = No.

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278 **Supplemental Table 3. List of antibodies and reagents**279 **Murine antibodies**

Antibodies	Source	Cat#
Anti-mouse PDCA1-BV421	BioLegend	127023
Anti-mouse CD4-FITC	BioLegend	100406
Anti-mouse CD21-APC	BioLegend	123412
Anti-mouse B220-PE-CY7	BioLegend	103222
Anti-mouse MHCII-BV510	BioLegend	107636
Anti-mouse CD11c-percp5.5	BioLegend	136504
Anti-mouse CD62L-BV510	BioLegend	104441
Anti-mouse CD11c-FITC	BioLegend	117306
Anti-mouse CD19-BV510	BioLegend	115546
Anti-mouse CD44-PE-CY7	BioLegend	103030
Anti-mouse CD23-FITC	BioLegend	101606
Anti-mouse CD3-APC	BioLegend	100236
Anti-mouse CD172a-APC	BioLegend	144014
Anti-mouse CD8 α -Percp-cy5.5	BioLegend	100736
Anti-mouse SiglecH-PE	eBioscience	12-0333-82
Total IRF7 antibody	Santa Cruz	sc-74471
Anti-EEA1	Abcam	ab2900
Goat Anti-Rabbit IgG H&L (Alexa Fluor® 647)	Abcam	ab150083

Donkey polyclonal Secondary Antibody to Goat IgG - H&L (Alexa Fluor® 488)	Abcam	ab150129
Goat polyclonal to NCF1/p47-phox	Abcam	ab166930
Rabbit polyclonal to RAC2	Abcam	ab191527
Alexa Fluor® 647 anti-mouse CD107a (LAMP-1) Antibody	Biolegend	121610
Alexa Fluor® 647 Goat anti-mouse IgG (minimal x-reactivity) Antibody	Biolegend	405322
Anti-mouse IFN γ -APC	BD	562018
Anti-mouse IL-17A-PE	BD	561020
DAPI	BD	564907
β -Actin (13E5) Rabbit mAb	Cell Signaling Technology	#4970
Anti-rabbit IgG, HRP-linked Antibody	Cell Signaling Technology	#7074
Anti-mouse IgG, HRP-linked Antibody	Cell Signaling Technology	#7076
Phospho-IRF7 (Ser437/438) (D6M2I) Rabbit mAb	Cell Signaling Technology	#24129
NF- κ B p65 (D14E12) XP® Rabbit mAb	Cell Signaling Technology	#8242
Phospho-NF- κ B p65 (Ser536) (93H1) Rabbit mAb	Cell Signaling Technology	#3033
Catalase (D5N7V) Rabbit mAb	Cell Signaling Technology	#14097
Anti-TLR7	Novus	NBP2-24906
Anti-TLR9	Novus	NBP2-24729
Phospho-p47phox (Ser370) Polyclonal Antibody	Thermo Fisher	PA5-36863

InVivoMAb anti-mouse CD317	Bio X Cell	BE0311
InVivoMAb rat IgG2b isotype control	Bio X Cell	BE0090

280

281 **Human antibodies**

Antibodies	Source	Cat#
Anti-human CD19-BV650	BioLegend	302238
Anti-human CD38-BV605	BioLegend	303532
Anti-human IgD-BV510	BioLegend	348220
Anti-human CD8-pacific blue	BioLegend	301033
Anti-human PD1-PE-CF594	BioLegend	329940
Anti-human CXCR3-PE	BioLegend	353705
APC anti-human Lineage Cocktail (CD3, CD14, CD16, CD19, CD20, CD56)	Biolegend	348803
Anti-human CD24-BV711	BD	563401
Anti-human CD11c-BUV395	BD	563787
Anti-human CD3-BV786	BD	563799
Anti-human CD4-BUV496	BD	564651
Anti-human CXCR5-AF647	BD	558113
Anti-human CD25-APC-R700	BD	565106
Anti-human CD127-BB700	BD	566398
Anti-human CD56-BUV737	BD	564447

Anti-human CD123-PECY7	BD	560826
Anti-human CD11c-percepcy5.5	BD	565227
Anti-human HLA-DR-FITC	BD	555811
Anti-human AXL-APC	R&D	FAB154A
Anti-human CD27-APC-EF780	eBioscience	47-0279
Anti-human CD45RA-PE-CY7	eBioscience	25-0458-73
Fc block	BioLegend	422301
LIVE/DEAD® Stain Kit Green Fluorescent	Invitrogen	L23101

282

283 **Reagents and kits**

Reagents	Source	Cat#
2-Deoxy-D-glucose (2-DG)	Selleck Chemical	S4701
Acetylcysteine (N-acetylcysteine)	Selleck Chemical	S1623
Etomoxir sodium salt	Selleck Chemical	S8244
Metformin HCl	Selleck Chemical	S1950
Hydroxychloroquine Sulfate	Selleck Chemical	S4430
Bafilomycin A1	Selleck Chemical	S1423
2-Guanidinobenzimidazole	Sigma	G11802
Cell Staining Buffer	BioLegend	420210
Recombinant Murine Flt3-Ligand	Peprtech	250-31L
Mouse IFN-alpha ELISA Kit	R&D	42120

Human IFN-alpha ELISA Kit	R&D	41100
Mouse IFN-beta ELISA Kit	R&D	42400
ELISA MAX™ Deluxe Set Mouse IL6	Biolegend	431304
ELISA MAX™ Deluxe Set Mouse TNF α	Biolegend	430904
R848 (Resiquimod)	Invivogen	tlrl-r848
ODN 2216	Invivogen	tlrl-2216
PtdIns-(3,4)-P2 (1,2-dihexanoyl) (sodium salt)	Cayman Chemical	10007759
1-Palmitoyl-3-oleoyl-sn-glycero-2-PE	Cayman Chemical	15104
1-Palmitoyl-2-oleoyl-sn-glycero-3-PC	Cayman Chemical	15102
LBIS Mouse IgG Rheumatoid Factor ELISA Kit	FUJIFILM Wako Shibayagi Corporation	637-02679
LBIS Mouse IgM Rheumatoid Factor ELISA Kit	FUJIFILM Wako Shibayagi Corporation	634-02689
LBIS Mouse anti-dsDNA ELISA Kit	FUJIFILM Wako Shibayagi Corporation	631-02699
OxyBURST™ Green H2DCFDA	Thermo Fisher	D2935
LysoSensor™ Green DND-189	Thermo Fisher	L7535
TaqMan™ Genotyping Master Mix	Thermo Fisher	4371353
MitoSOX™ Red	Thermo Fisher	M36008
SuperBlock™ (TBS) Blocking Buffer	Thermo Fisher	37535
Anti-Mouse/Rat Foxp3 Staining Set PE	eBioscience	72-5775-40

BD™ Cytometric Bead Array (CBA) Mouse TNF Flex Set	BD	558299
BD™ Cytometric Bead Array (CBA) Mouse IL6 Flex Set	BD	558301
Fixation/Permeabilization Solution Kit with BD GolgiStop™	BD	554715
Long Range PCR kit	Qiagen	206402
Diamond Plasmacytoid Dendritic Cell Isolation Kit II, human	Miltenyi Biotech	130-097-240
pLKO.1-shRac2-puro-CMV-tGFP lentivirus	Sigma	TRCN0000065343

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296 **Supplemental Table 4. Primers used in this study.**

Genes	Forward primer (5'-3')	Reverse primer (5'-3')
mouse mx1	CTGAGGGCTCTGGGTGT	GTAACAATACCACTGCCTCTG
mouse irf7	CCTGATCCTGGTGAAGCTGG	TGGGAGTTGGGATTCTGAGTC
mouse oas1	TTTGAGCAGGTAGAAGAGAACT	GCATCAGAAGCACGGAGT
mouse isg15	AGAGCAAGCAGCCAGAAG	CACCGTCATGGAGTTAGTCAC
mouse ifit1	ATGGGAGAGAATGCTGATGG	AGGAACTGGACCTGCTCTGA
mouse β -actin	ATGCTCCCCGGGCTGTAT	CATAGGAGTCCTTCTGACCCATTC
mouse Catalase clone	GGAATTCATGTCGGACAGTCGGGAC C	GGCGGCCGCTTACAGGTTAGCTTTT CCCTTC
human mx1	GGGTAGCCACTGGACTGA	AGGTGGAGCGATTCTGAG
human ifit1	GCCTCCTTGGGTTTCGTCTACAA	TCAAAGTCAGCAGCCAGTCTCA
human oas1	GAAGGCAGCTCACGAAAC	TTCTTAAAGCATGGGTAATTC
human irf7	TGAAGCTGGAACCCTGG	GATGTCGTCATAGAGGCTGTT
Human rpl13a	CCTGGAGGAGAAGAGGAAAGAGA	TTGAGGACCTCTGTGTATTTGTCAA

297

298 **Taqman assay for *NCF1* genotyping**

	Forward primer	Reverse primer	Reporter 1 Sequence	Reporter 2 Sequence
NCF1 p. R90H	CAGCTCCCAAG TGGTTTGAC	GGTGGGCA GGCTCATGA	CCTGGCGGTTCT C	CCTGGTGGTT CTC

299