

## Supplemental Information

### Supplemental Figure Legends

#### Figure S1

Flow cytometric analysis of pSTAT1 in peripheral CD3<sup>+</sup> T cells, CD19<sup>+</sup> B cells, and CD14<sup>+</sup> monocyte following to IFN- $\gamma$  stimulation. The representative histogram obtained from in healthy control and patient with CMCD. Black line: no stimulation. Blue line: IFN- $\gamma$  for 15 min. The MFI of the pSTAT1 signal is indicated in the figure.

#### Figure S2

Flow cytometric analysis of pSTAT1 on 0 and 1 day after blood collection in peripheral CD14<sup>+</sup> cells with or without staurosporine treatment following to IFN- $\gamma$  stimulation. The representative histogram obtained from healthy control. Black line: no stimulation. Blue line: IFN- $\gamma$  for 15 min. Magenta lines: incubation with staurosporine for 15 min after IFN- $\gamma$  stimulation.

#### Figure S3

Impaired dephosphorylation of STAT1 in the cells of patients, compared to dephosphorylation observed in cells of healthy controls. (A) The representative histogram obtained from P1 and healthy control. Black line: no stimulation. Blue line: IFN- $\gamma$  for 15

min. Red and orange lines: incubation with staurosporine for 15 (red) or 30 (orange) min after IFN- $\gamma$  stimulation. (B) The percentage decrease in the amount of pSTAT1 in the presence of staurosporine, relative to the total amount of pSTAT1 detected after IFN- $\gamma$  stimulation alone, was calculated by determining the delta MFI for these two conditions for 6 patients and 14 healthy individuals, as described (b/a x 100). CD14<sup>+</sup> cells from the patient displayed a significantly smaller decrease in pSTAT1 levels in the presence of staurosporine, than CD14<sup>+</sup> cells from healthy controls under the same conditions (\* $p$ <0.02).

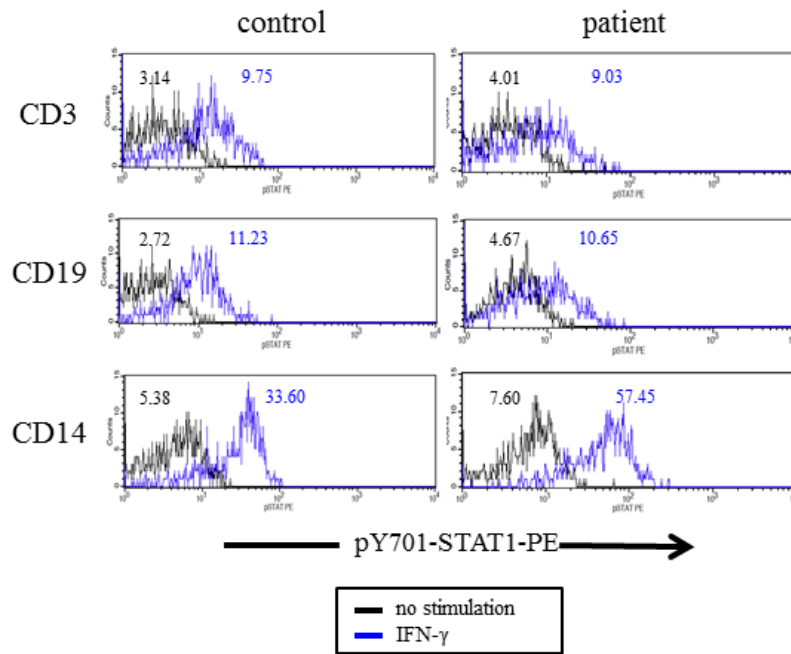
#### **Figure S4**

The level of pSTAT1 after IFN- $\gamma$  stimulation obtained in Fig 4A was quantified by densitometry. The data are expressed as fold-induction relative to WT cells, which is set to 1.

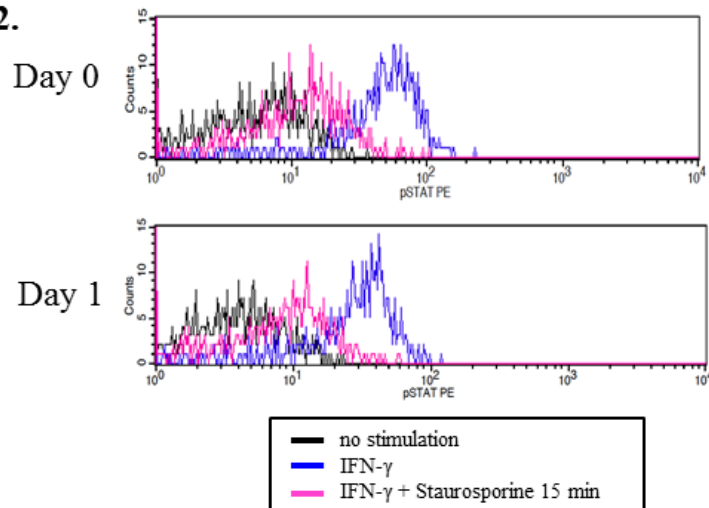
#### **Figure S5**

GAS reporter assay in response to IL-27 treatment. IL-27 strongly induced GAS activation in all of the CMCD-related mutations. The data are expressed as fold-inductions relative to WT cells, set to 1. \*Differences were statistically significant in the cells carrying the mutants compared with the cells carrying WT ( $P$ <0.05).

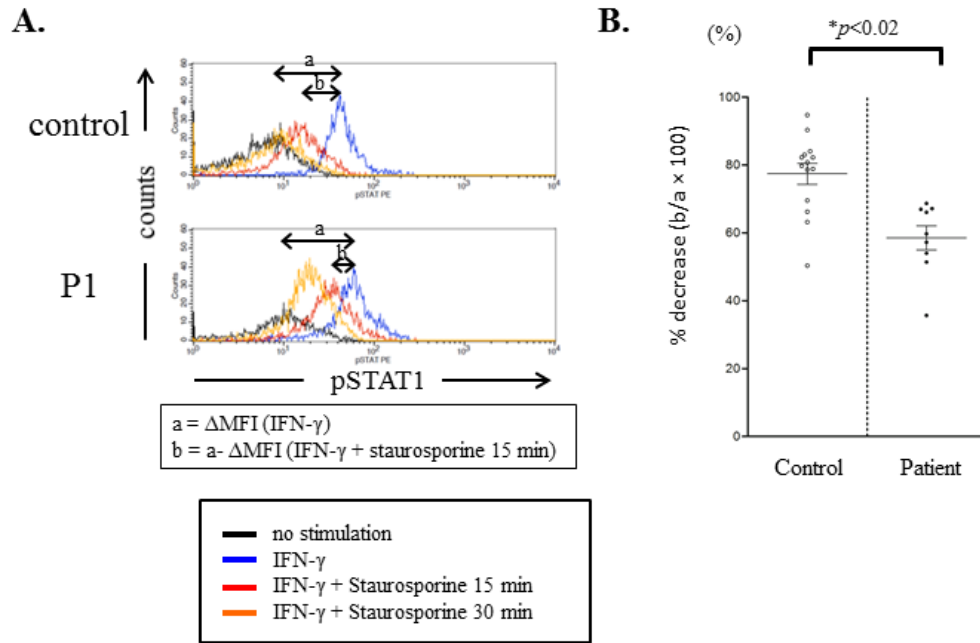
**Figure S1.**



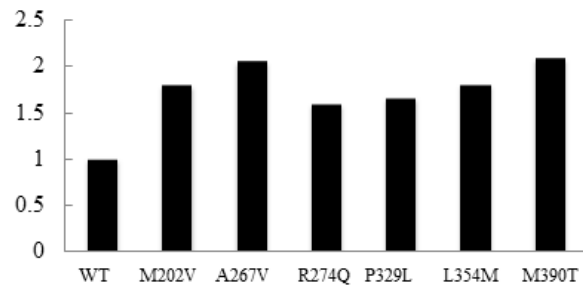
**Figure S2.**



**Figure S3.**



**Figure S4.**



**Figure S5.**

