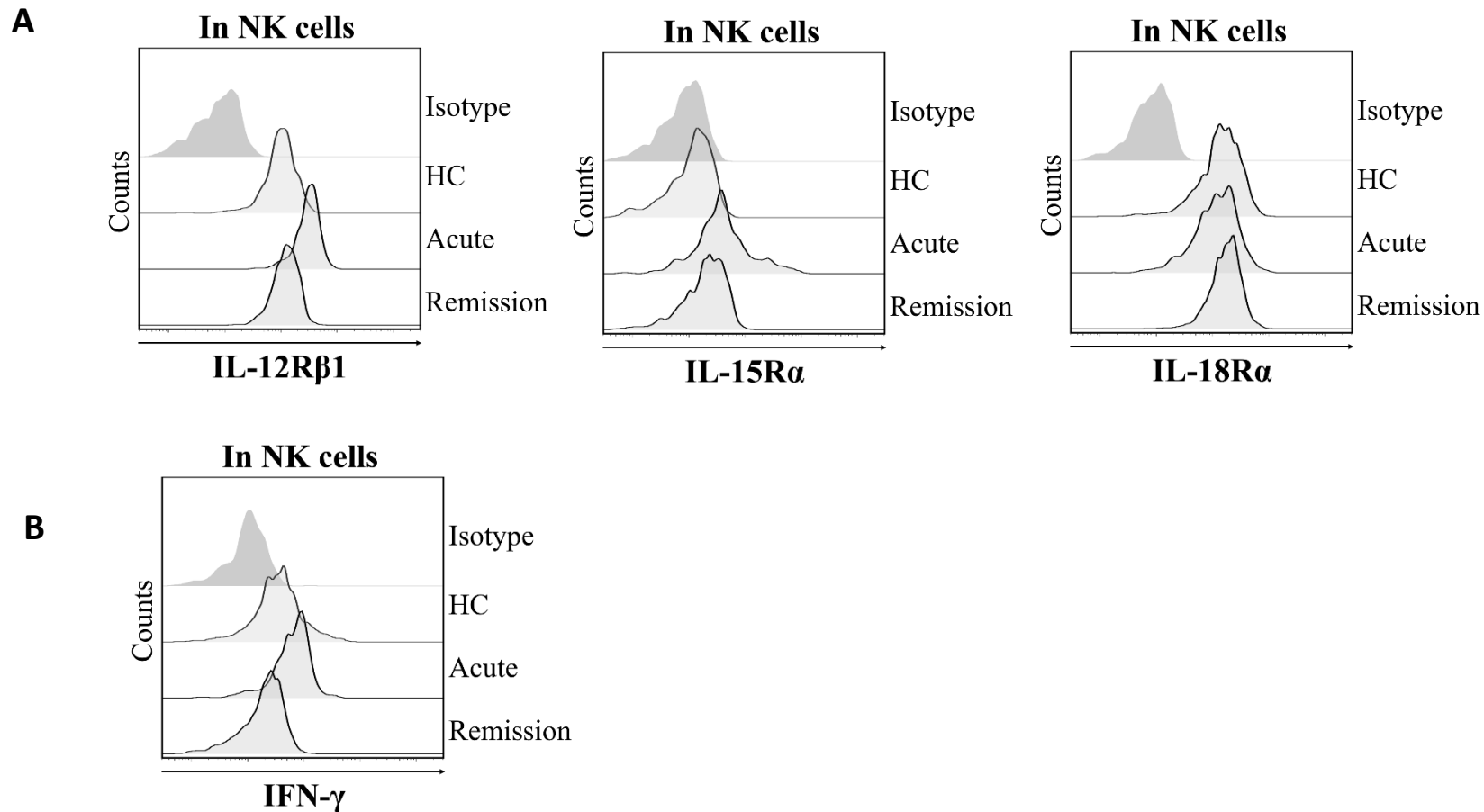
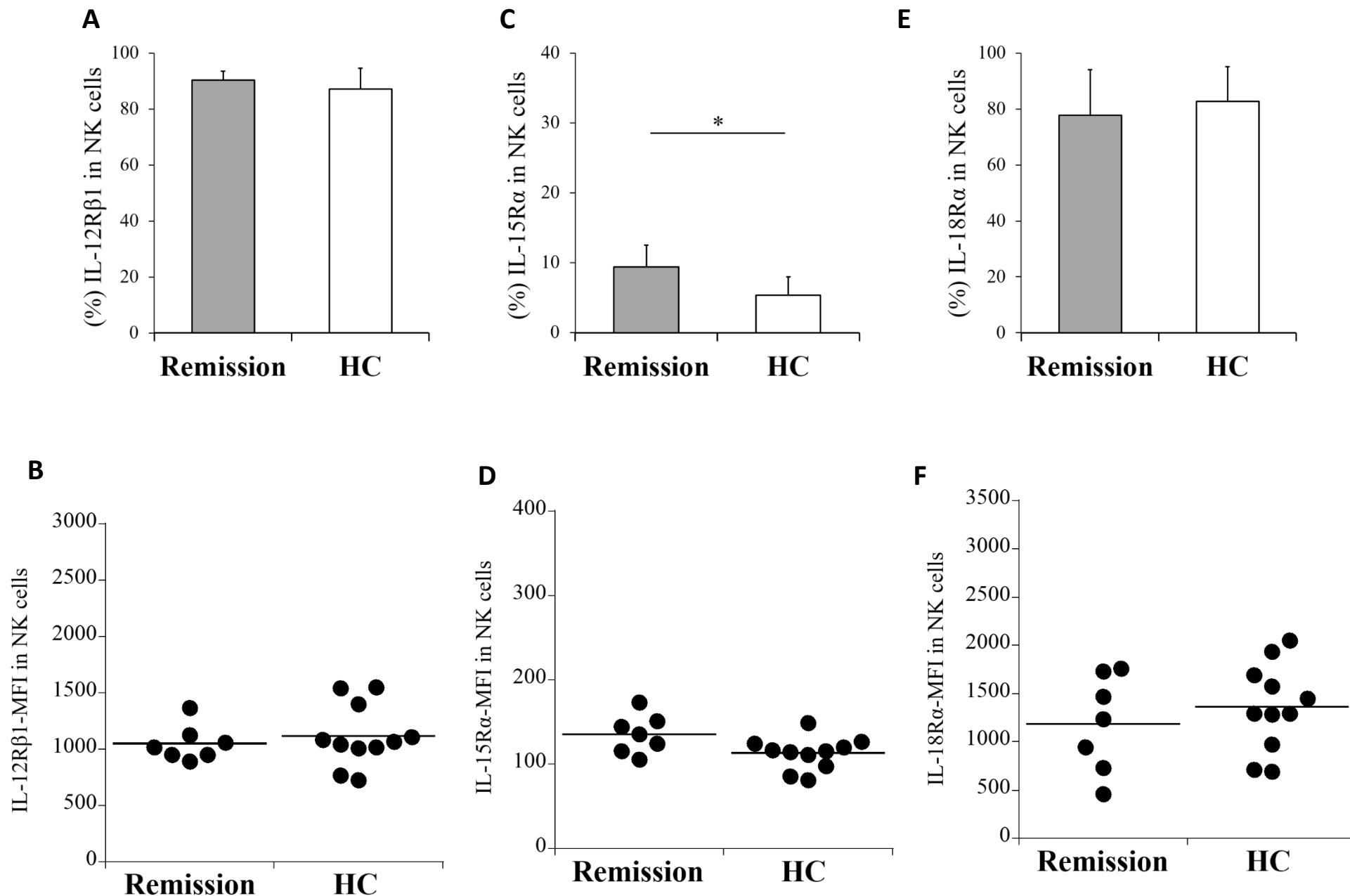


## Supplementary Figure 1



**Expression of interleukin (IL)-12R $\beta$ 1, IL-15R $\alpha$ , or IL-18R $\alpha$  on NK cells, and interferon (IFN)- $\gamma$  in NK cells**  
The histograms indicate the representative findings of IL-12R $\beta$ 1, IL-15R $\alpha$ , or IL-18R $\alpha$  expression on NK cells (**A**), and IFN- $\gamma$  expression in NK cells (**B**) from patient with acute AOSD, patient with remission AOSD, and healthy control (HC).

## Supplementary Figure 2



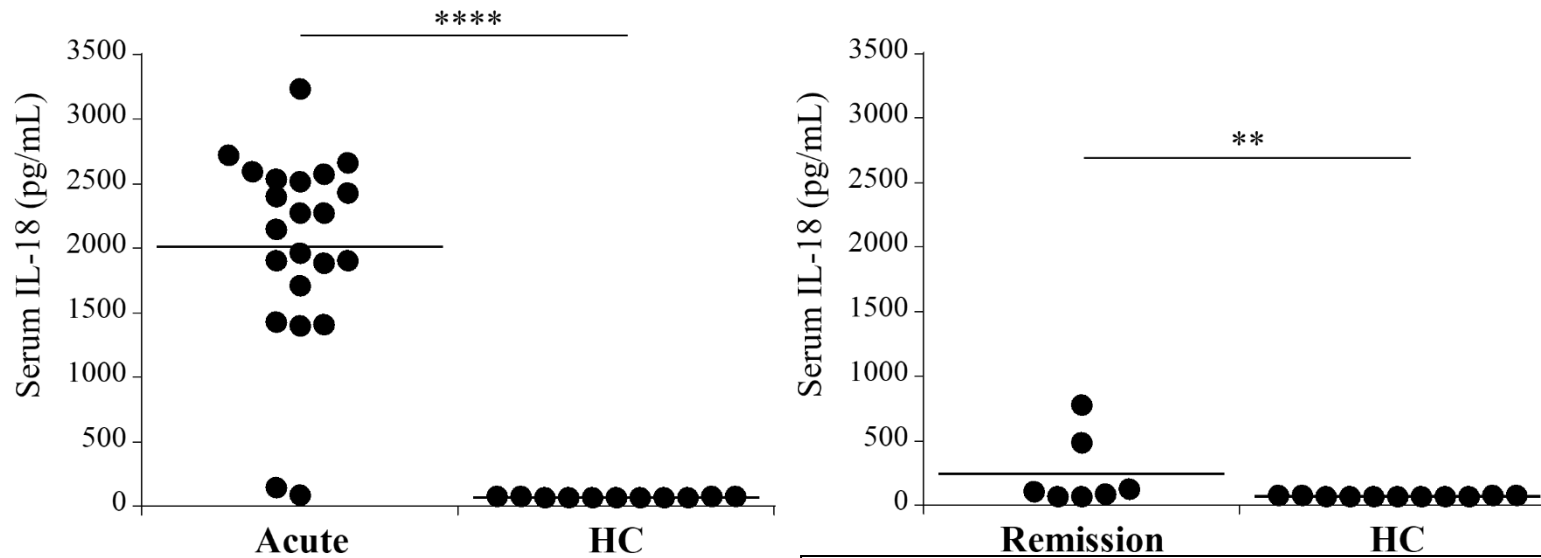
**Supplementary Figure 2 Legend:**

**The comparison of IL-12R $\beta$ 1, IL-15R $\alpha$ , and IL-18R $\alpha$  expression on NK cells between remission AOSD and HC**

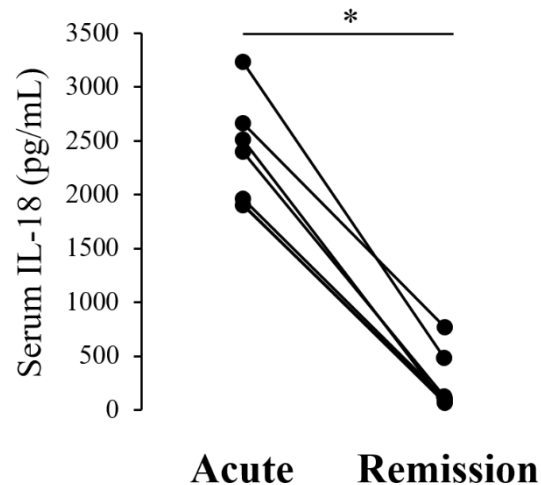
Percentage of IL-12R $\beta$ 1, IL-15R $\alpha$ , or IL-18R $\alpha$  (**A, C, E**) and median fluorescence intensity (MFI) of them (**B, D, F**) in NK cells were compared between patients with remission AOSD and HC. Values are shown as the mean  $\pm$  SD. The Mann-Whitney U test was employed in the comparison. Statistically significant difference is indicated as  $^*P < 0.05$ .

### Supplementary Figure 3

**A**

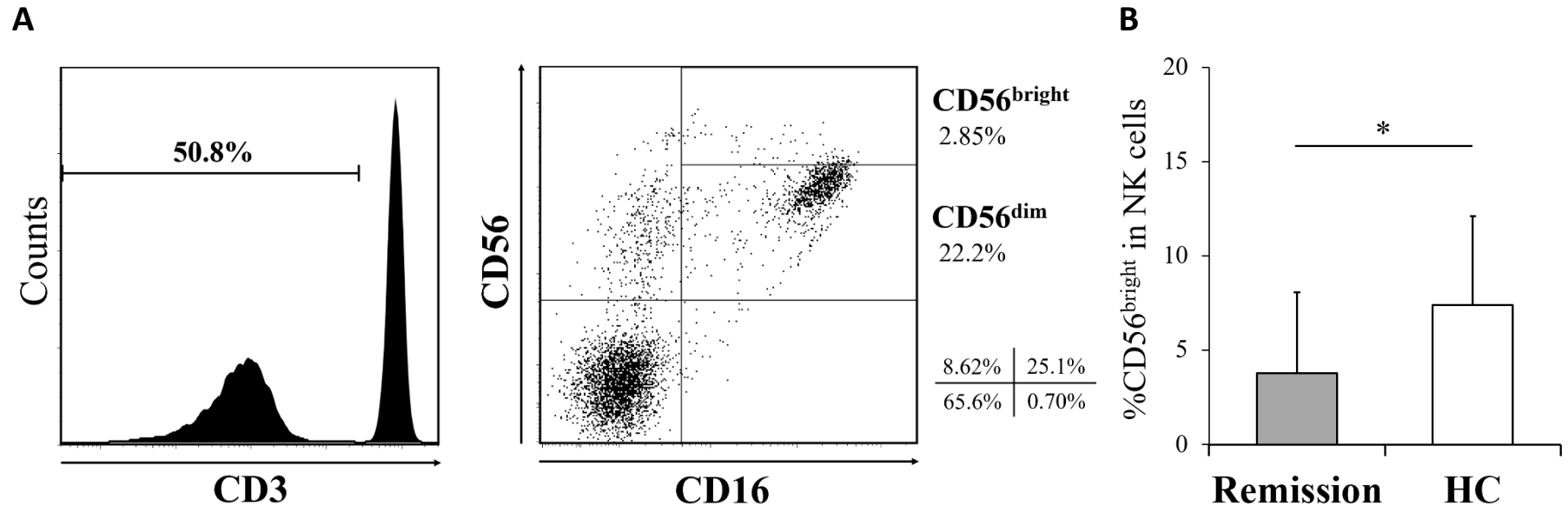


**B**



Serum IL-18 levels were compared in patient with acute AOSD, those with remission AOSD, and HC (A). They were sequentially evaluated in 7 patients between acute and remission phase (B). Values are shown as the mean  $\pm$  SD. The Mann-Whitney U test was employed in the comparison between three groups. The Wilcoxon signed-ranked test was used in that between an acute and remission phase. Statistically significant differences are indicated as follows: \* $P < 0.05$ , \*\* $P < 0.01$ , and \*\*\*\* $P < 0.0001$ .

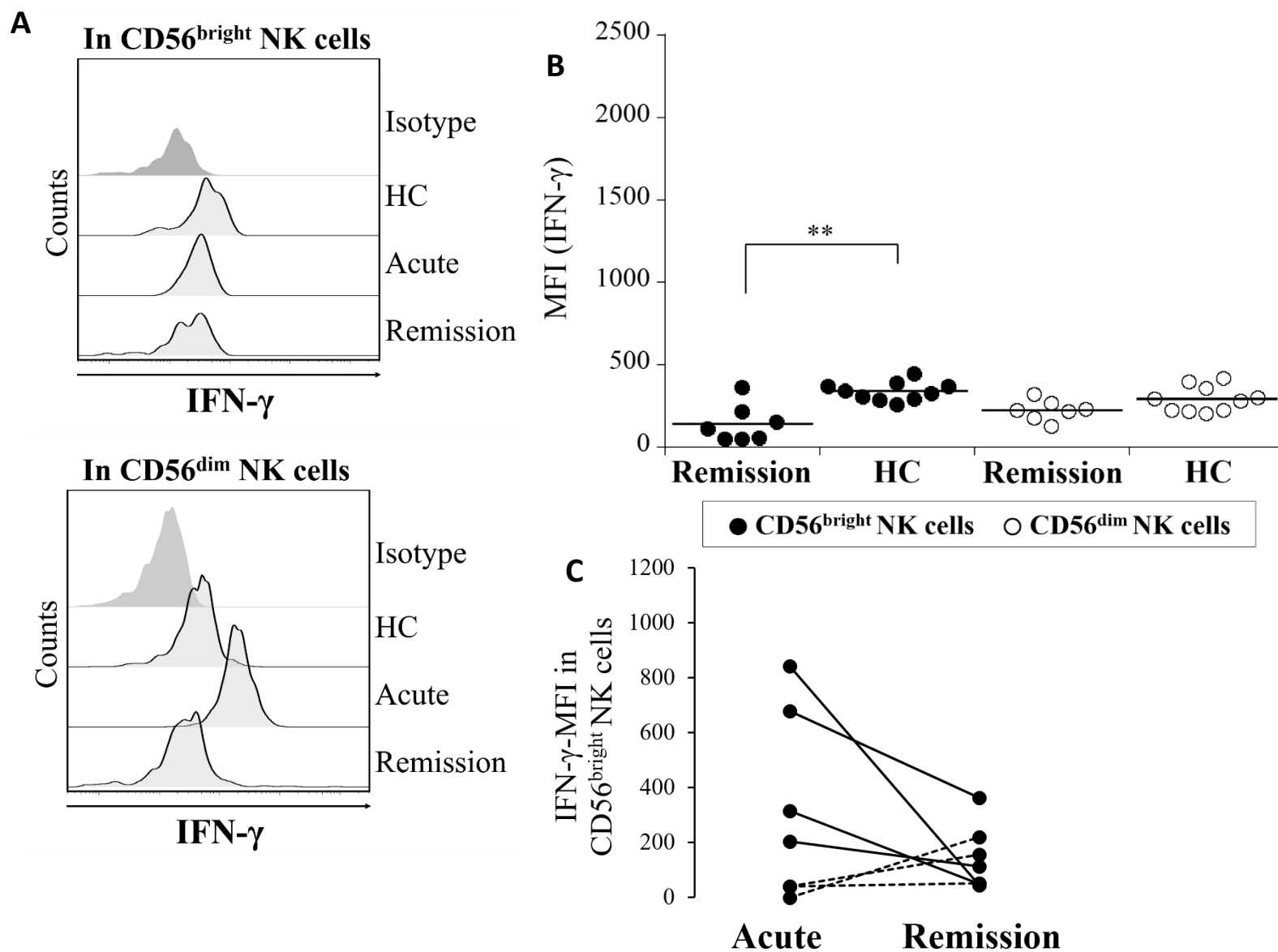
## Supplementary Figure 4



### The proportion of CD56<sup>bright</sup> or CD56<sup>dim</sup> NK cells in two distinct NK cell subsets.

The representative histogram of CD3 negative population in total lymphocytes and dot plot of CD16/CD56<sup>bright/dim</sup> expression in CD3 negative population were displayed (A). The proportion of CD56<sup>bright</sup> in NK cells was compared between patients with remission AOSD and HC (B). Values are shown as the mean  $\pm$  SD. The Mann-Whitney U test was employed in the comparison. Statistically significant difference is indicated as \* $P < 0.05$ .

**Supplementary  
Figure 5**



**Supplementary Figure 5 Legend:**

**IFN- $\gamma$ -producing ability in two distinct NK cell subsets**

In the population of CD56<sup>bright</sup> or CD56<sup>dim</sup> NK cells, IFN- $\gamma$ -producing ability was evaluated. The representative histograms of that were shown (A). IFN- $\gamma$ -MFI in each subset was compared between patients with remission AOSD and HC (B). IFN- $\gamma$ -MFI in CD56<sup>bright</sup> NK cell population was compared between an acute and remission phase; the dotted lines are used for showing patients having the treatment with biologics (C). Values are shown as the mean  $\pm$  SD. The Mann-Whitney U test was employed in the comparison between patients with remission AOSD and HC. The Wilcoxon signed-ranked test was used for the comparison between CD56<sup>bright</sup> and CD56<sup>dim</sup> in remission AOSD, or for that between an acute and remission phase. Statistically significant difference is indicated as <sup>\*\*</sup> $P < 0.005$ .