

## Supplementary information

**Supplementary table.** Demographics and baseline disease characteristics

| Variables                    | Patients<br>(n = 35) |
|------------------------------|----------------------|
| Age, years                   |                      |
| Mean $\pm$ SD                | 34.0 $\pm$ 14.9      |
| Median (range)               | 36.0 (9–74)          |
| Female, n (%)                | 25 (71.4)            |
| Caucasians, n (%)            | 33 (94.3)            |
| Form of CAPS, n (%)          |                      |
| MWS                          | 33 (94.3)            |
| MWS/NOMID                    | 4 (11.4)             |
| <i>NLRP3</i> mutation, n (%) |                      |
| R260W                        | 18 (51.4)            |
| T348M                        | 7 (20.0)             |
| D303N                        | 3 (8.6)              |
| E311K                        | 2 (5.7)              |
| Other*                       | 5 (14.3)             |
| Previous treatment, n (%)    |                      |
| Canakinumab                  | 9 (25.7)             |
| Anakinra                     | 17 (48.6)            |

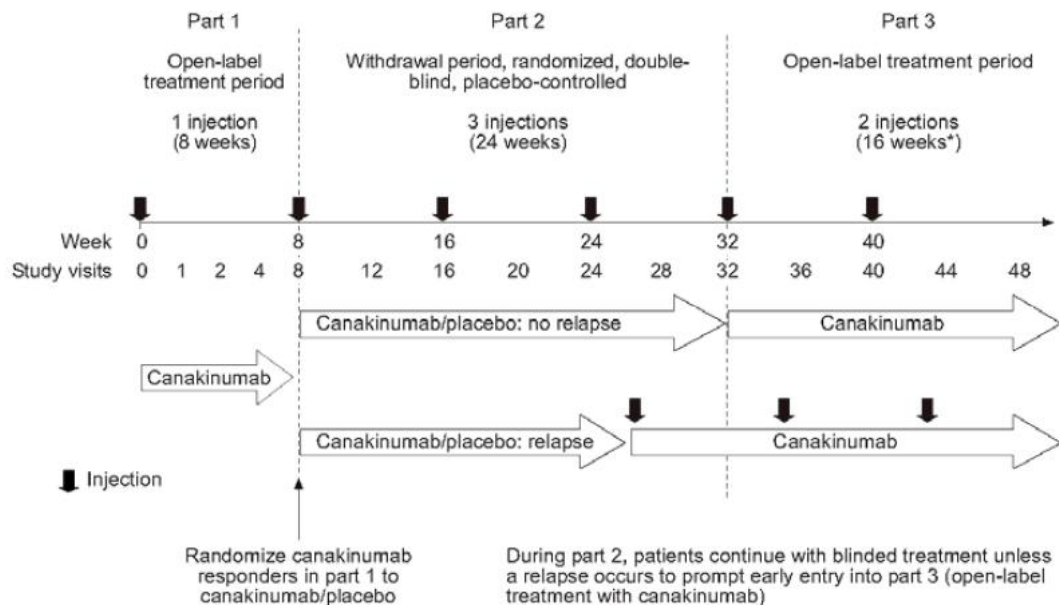
\*One each of M662T, A439V, D305N, T436N, and T436I.

CAPS, cryopyrin-associated periodic syndrome; MWS, Muckle–Wells syndrome; NOMID, neonatal-onset multisystem inflammatory disease.

## Supplementary figure 1. Study design

The time point for entering part 3 was based on completion of part 2 or relapse.

\*For patients who completed part 2 and part 3 had a duration of 16 weeks, with patients receiving two injections of canakinumab. For patients who relapsed in part 2 and part 3 had a duration of up to 40 weeks, with patients receiving canakinumab injections every 8 weeks.



## Supplementary figure 2. Patient disposition

CR, complete response; ITT, intent-to-treat.

\* one patient relapsed on the last day of part 2

