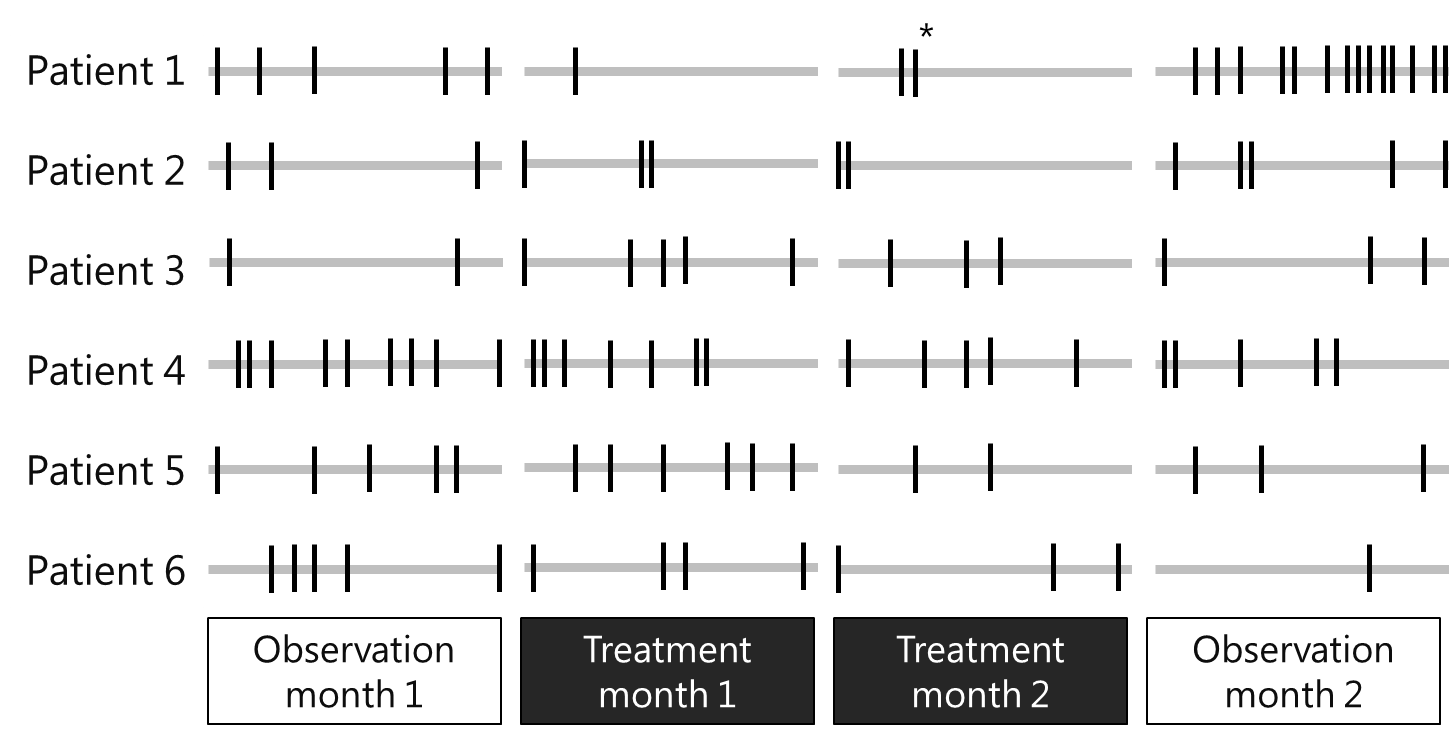
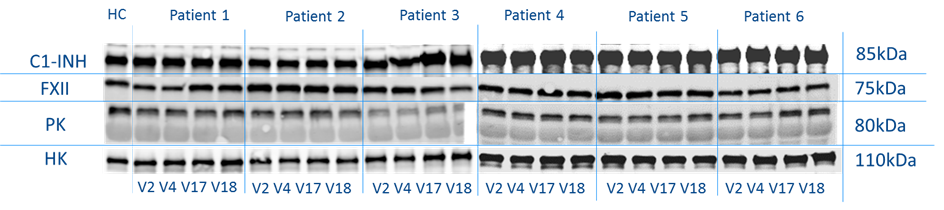
**FIGURES & SUPPLEMENTAL FIGURES**



**Figure 1. Attack episodes per month**.

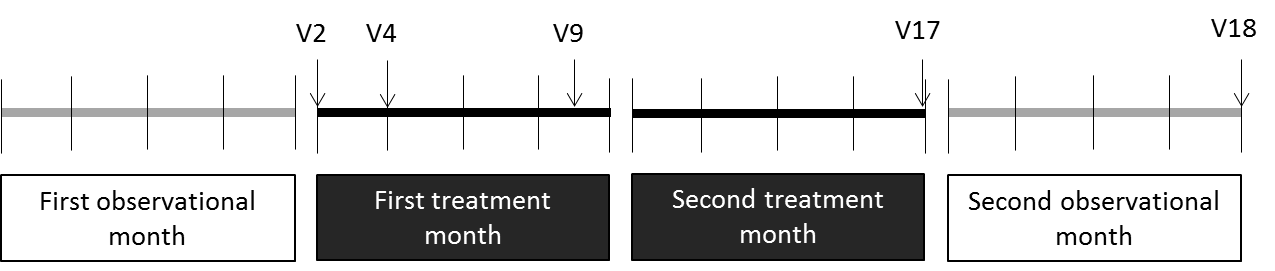
**Legend**: One attack episode is indicated with a vertical line and could last for multiple days. Attacks in the observation month versus the treatment month: Patient 1: 19 vs 3. Patient 2: 8 vs 5. Patient 3: 5 vs 8. Patient 4: 14 vs 12. Patient 5: 8 vs 8. Patient 6: 6 vs 7.

\* AE attack occurred seven days after last rhC1-INH administration due to a missed treatment visit



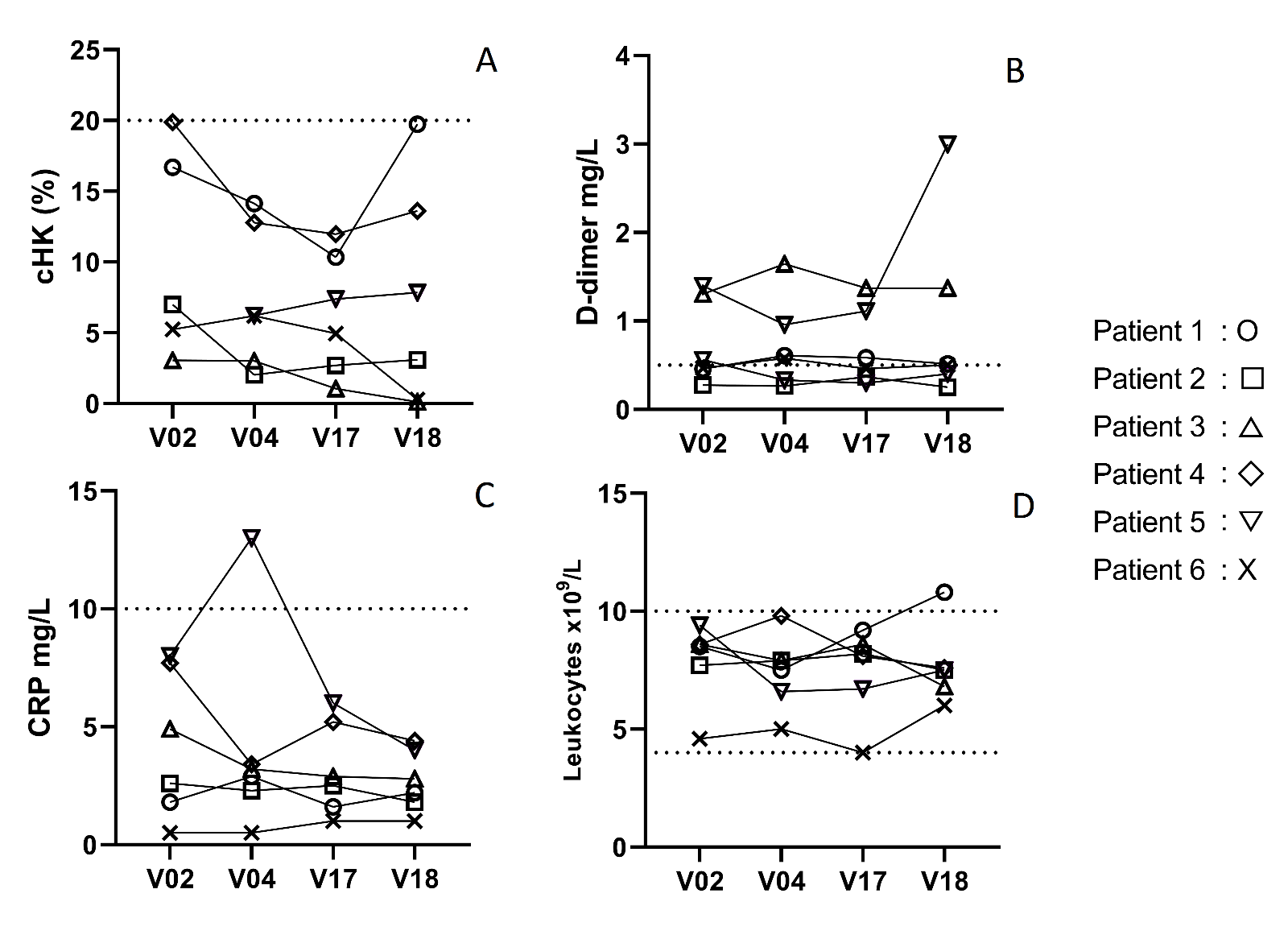
**Figure 2. Proteins of the contact system.**

**Legend**: Immunoblot of the proteins of the contact system C1-INH (C1-esterase inhibitor), HK (high molecular weight kininogen), PK (plasma kallikrein) and FXII (factor XII). Healthy Control (HC) represents a healthy pooled plasma sample. Visits 4 and 17 - samples were collected during the treatment visit, prior to administrating rhC1-INH. Visits 2 and 18 - samples were collected at the end of the two observation months.



**Supplemental figure 1. Trial design with blood drawn visits.**

**Legend**: Four month study sequence divided into weeks by vertical lines. Trial visits at which blood is drawn are indicated with an arrow. Visits occur twice a week during the treatment months in which rhC1-INH is administered. Visit 18 marks the end of the trial, no pdC1-INH was administered during this visit.

**Supplemental figure 2. Laboratory assessment**

**A) % Cleaved kininogen (cHK).** 100% indicates cleavage of the total HK pool**.** Visit 2 sample was collected prior to the first recombinant human C1-esterase inhibitor (rhC1-INH), visits 4 and 17 prior to third and sixteenth gift, visit 18 sample at final visit. Dotted lines represent upper range or normal ranges. Data V02 Patient 5 missing. Reference range <20% **B)** **D-dimer levels in mg/L over time.** Reference range 0.00-0.50 mg/L. **C) CRP (mg/L) over time during study.** Reference range <10mg/L. **D) Leukocytes (x109/L) over time during study**. Reference range 4.0-10.0 x109/L