

Supplemental Materials

Fig. S1. Thoracic computed tomographic images (CT).

No emboli were detected on Day -11 (**A**) and Day 15 (**B**).

Fig. S2. JCRG's analyses of the patient's FXIII and anti-FXIII antibodies.

A. A 5-step dilution cross-mixing test by an amine incorporation assay for FXIII activity. The mixed samples showed an 'inhibitor' pattern, because there was a downward deviation (arrow). A broken line depicts a theoretical 'deficient' pattern.

B. Immuno-blot assays were performed using rFXIII-A, rFXIII-B, and their complexes (A_2B_2) at the indicated amounts shown as antigen (ng). The results showed the presence of anti-FXIII-A antibodies in patient plasma.

C. A fibrin cross-linking study. The results showed both markedly delayed γ -dimerization (γ - γ) and the lack of α -polymerization reactions (α -poly).

D. An immuno-chromatographic test (ICT) to detect FXIII-A-bound anti-FXIII-A autoantibodies with (spiked) or without (direct) pre-mixing pooled normal plasma. From the left to the right stripes; direct ICT for negative control (the plasma obtained from a healthy control), direct and spiked ICTs for the present AH13 case (Patient), and spiked ICT for positive control (the plasma obtained from a previously diagnosed AH13 case).

Fig. S3. Pharmacokinetic (PK) studies by exogenous FXIII administration.

Bleeding events occurred three times in the first two months, and were successfully managed by administering large amounts of plasma-derived FXIII concentrates (gray arrows): 1,440U (34 U/kg) on Day 4 (**A**), and 2,400U (57 U/kg) on Day 18 (**B**), Day 53 (**C**) and Day 302 (**D**). Blood samples were taken at indicated time intervals (prior to infusion, 0.5h, 1h, 2h, 4h, 8h, 12h, 24h after infusion), and FXIII activity (filled circles) and antigen (filled squares) were measured.