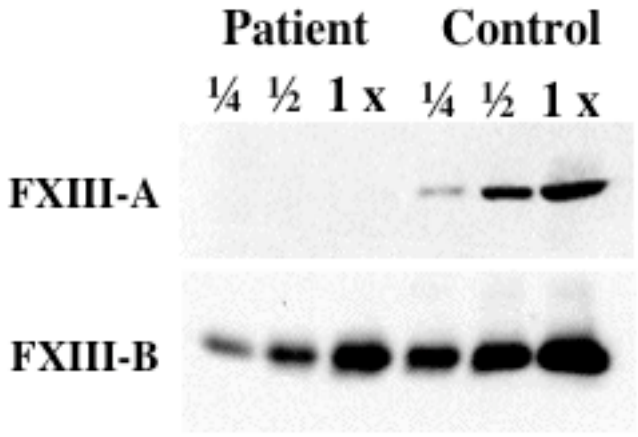
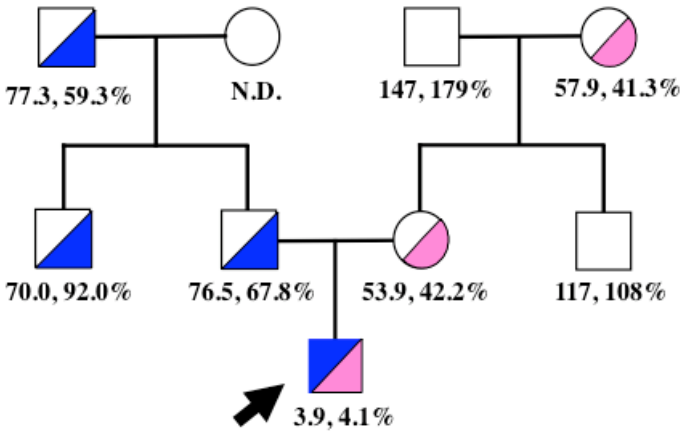
Supplementary Material to Fujii et al. "A short half-life of the administered factor XIII (FXIII) concentrates after the first replacement therapy in a newborn with severe congenital FXIII deficiency" (Thromb Haemost 2012; 107.3)



Suppl. Figure 1: FXIII-A and FXIII-B antigens in the proband's plasma. Essentially no FXIII-A antigen was detected by Western blotting, while FXIII-B protein was found to have decreased mildly in the patient (left) when compared to a normal control (right). An anti-FXIII-A antibody was rabbit polyclonal and homemade as described previously (Ichinose et al, Biochemistry 25; 4633-4638, 1986). An anti-FXIII-B antibody (RAHu/FXIII-S; polyclonal rabbit antiserum) was purchased from NORDIC immunological Laboratories (Tilburg, The Netherlands).



Suppl. Figure 2: Family pedigree and FXIII activity and FXIII-A antigen levels.
An arrow indicates the proband of severe FXIII deficiency in this family. FXIII activities and antigen levels are determined by an amine incorporation assay and ELISA, respectively. FXIII activities (% of normal) are followed by FXIII antigen levels (% of normal). Blue-fills and pink-fills stand for Tyr204Stop and Ser708Arg mutations, respectively. N.D.; not determined because a sample was not available.

Suppl. Table 1: Laboratory tests (day 5).

WBC	9,550	/μL	TP	4.5	g/dL
RBC	341×10^4	/μL	T-bil	12.4	mg/dL
Hb	11.8	g/dL	GOT	23	IU/L
Ht	34.7	%	GPT	7	IU/L
Plt	22.5×10^4	/μL	LDH	358	IU/L
			CK	129	IU/L
PT	9.9	sec	BUN	3	mg/dL
PT-INR	0.90		Cre	0.40	mg/dL
aPTT	48.6	sec	Na	145	mEq/L
Fibrinogen	121	mg/dL	K	4.2	mEq/L
FDP	3.1	μg/mL	Ca	9.0	mg/dL
Antithrombin	54.6	%			
F□/8 act.	104.6	%	CRP	0.02	mg/dL
F□/9 act.	25.7	%			
vWF	173	%			