Study definitions:

Moderate von Willebrand disease (VWD) is defined as haemorrhagic symptoms or a family history of VWD and von Willebrand factor (VWF) antigen (VWF:Ag), VWF ristocetin co-factor activity (VWF:RCo) or VWF collagen binding activity (VWF:CB) 11–30 IU/dL and/or factor VIII levels (FVIII:C) ≤ 40 IU/dL in the absence of haemophilia A.

Severe VWD is defined as haemorrhagic symptoms or a family history of VWD and VWF antigen (VWF:Ag), VWF ristocetin co-factor activity (VWF:RCo) or VWF collagen binding activity (VWF:CB) ≤ 10 IU/dL and/or factor VIII levels (FVIII:C) ≤ 10 IU/dL in the absence of haemophilia A.

Type 3 VWD is defined as VWF:Ag and VWF:Act < 5 IU/dL.

Moderate haemophilia A is defined as FVIII:C or FIX:C 1–5 IU/dL.

Severe haemophilia A is defined as FVIII:C or FIX:C < 1 IU/dL.

A verified joint bleed is defined as any joint symptom requiring treatment with DDAVP or coagulation factor at least once, as documented in the medical file; this definition has been the same across all previous studies from which data were extracted for the current study.

Prophylaxis is defined as any history of at least 1 regular clotting factor concentrate infusion per week for at least 45 consecutive weeks. This definition has been the same across all previous studies from which data were extracted for the current study.

Home treatment is defined as self-infusion at home of coagulation factor concentrates on the basis of on demand or prophylactic treatment (same definition across studies).

Clinically significant inhibitor: FVIII inhibitor: a positive inhibitor titer (more than 0.6 Bethesda Unit/mL) and decreased recovery after FVIII infusion. For VWD patients as documented in the medical files based on decreased recovery and clinical response after clotting factor infusion.