

## APPENDIX 2: Treatment options

International Non-proprietary Name / Generic Name	Product	Formulation	Properties	Adverse reactions
Desmopressin (DDAVP)	Octostim	Solution for injection i.v., s.c. (15 micrograms/ml) Nasal spray i.n. (150 micrograms/dose)	Synthetic analogue of vasopressin. Release of endogenous FVIII, VWF and tissue plasminogen activator (t-PA).	Headache, nausea, temporary lowering of blood pressure with secondary tachycardia, facial flushing. Fluid retention, hyponatremia and seizures.
Factor VIII/VWF	Haemate Haemate - P	Sterile, lyophilized powder for injection i.v. (1000 IE FVIII, 2400 IE VWF), (500 IE FVIII, 1200 IE VWF)	Plasma-derived factor concentrate. Pasteurization 10 h at 60°C. VWF:RC <sub>0</sub> /FVIII:C approx. 2.7	Theoretical risk of prion transmission. Allergic / anaphylactic reactions. Development of alloantibodies.
	Wilate	Sterile, lyophilized powder for injection i.v. (900 IE FVIII, 800 IE VWF), (450 IE FVIII, 400 IE VWF)	Plasma-derived factor concentrate. Solvent-detergent treatment, heating of the lyophilized product for 2 h at 100°C (PermaHeat). VWF:RC <sub>0</sub> /FVIII:C approx. 0.8	Theoretical risk of prion transmission. Allergic / anaphylactic reactions. Development of alloantibodies.
VWF	Wilfactin Willfact	Sterile, lyophilized powder for injection i.v. (10 IE FVIII, 1000 IE VWF)	Plasma-derived. Solvent-detergent treatment, 35 nm nanofiltration and dry heating of the lyophilized product at 80°C for 72 h. VWF:RC <sub>0</sub> /FVIII:C approx. 60 Priming dose of FVIII may be necessary.	Theoretical risk of prion transmission. Allergic / anaphylactic reactions. Development of alloantibodies.
Tranexamic acid	Cyklokapron, Cyklonova	Solution for injection i.v. Tablets Dissolvable tablets Mouthwash	Anti-fibrinolytic	Nausea, vomiting, diarrhoea and abdominal pain
Hormonal therapy	Estrogen containing oral contraceptives  Intrauterine device	Tablet	Variable increase of VWF  Progesteron induced reduction in bleed	Headache, nausea, weight gain, thrombosis, cancer Headache, weight gain, risk for infection
Transfusion	Platelets	Infusion		Allergic reactions, development of alloantibodies.