

### Appendix 3.

Treatment with VWF/FVIII or VWF concentrates at different clinical situations.

Clinical situation	Dose of VWF:RCo <sup>1</sup>	No. of infusions	VWF:RCo target plasma level <sup>2</sup>
Major surgery	40 – 60 IU/kg	Every 12 h initially, then once daily until wound healing is complete.	0.50 – 1.00 kIU/L; maintain levels for 3 – 10 days
Minor surgery	30 – 50 IU/kg	Once daily (may require only 1 – 3 days).	> 0.30 kIU/L
Dental extractions	20 – 30 IU/kg	Usually one dose inly before procedure.	> 0.30 kIU/L for >12 h
Spontaneous bleeding	20 – 60 IU/kg	One daily dose until bleeding stops. Monitor clinically.	>0.30 kIU/L
Delivery and puerperium	50 IU/kg	Once daily.	>0.50 kIU/L; maintain levels for 3 – 4 days

Table modified from: Mannucci PM and Franchini M In Von Willebrand Disease. Basic and Clinical aspects (Federici AB, Lee CA, Berntorp E, Lillicrap D and Montgomery RR, eds.). Wiley-Blackwell 2011, p. 203.

Footnotes:

- 1) The dosages are indicated for patients with VWD with severely reduced FVIII:C and/or VWF:RCo levels (<0.10 kIU/L).
- 2) Factor levels can be predicted based on pharmacokinetic data; 40-50 units/kg (VWF:RCo) will increase plasma VWF levels to 80-100 % depending on the baseline levels and haematocrit.