

Supplementary Material

The international prospective Glanzmann Thrombasthenia Registry: treatment and outcomes in surgical intervention

Supplementary Table 1. Dosages, dose schedule and treatment duration for surgical episodes treated with rFVIIa (alone or in association with other hemostatic agent[s]).

Patient group/ surgical category	Dosage ($\mu\text{g}/\text{kg}$)		Number of doses		Interval between doses (h)		Cumulative dose ($\mu\text{g}/\text{kg}$)		Duration of treatment (h)	
	Mean \pm SD	Median IQR Range	Mean \pm SD	Median IQR Range	Mean \pm SD	Median IQR Range	Mean \pm SD	Median IQR Range	Mean \pm SD	Median IQR Range
<i>A. Minor procedures (n=169)</i>										
No	101.5	90	2.8	2.0	13.3	2.0	398.4	180	30.3	4.0
AB/refractoriness (n=44)	\pm 34.7	90.0-90.0 37.0-180.0	\pm 2.9	2.0-3.0 1.0-20.0	\pm 87.3	2.0-3.0 2.0-749.0	\pm 1,260. 5	180.0-270.0 80.0-8,544.0	\pm 131.4	3.0-6.0 2.0-749.0
AB+refractoriness (n=40)	124.6 \pm 40.7	102 95.0-160.0 70.0-250.0	3.0 \pm 1.6	3.0 2.0-3.0 1.0-7.0	4.2 \pm 4.8	2.0 2.0-3.0 2.0-26.0	373.8 \pm 232.1	300 193.2-498.0 85.0-1,250.0	9.6 \pm 16.4	4.0 2.0-6.0 2.0-68.0
Refractoriness only (n=11)	144.7 \pm 49.9	117 100.0- 200.0 85.0-200.0	4.3 \pm 5.3	3.0 2.0-4.0 1.0-20.0	2.9 \pm 1.8	2.0 2.0-3.5 2.0-12.0	674.3 \pm 1,313. 7	300 170.0-360.0 90.0-4,617.0	11.7 \pm 16.1	6.0 5.0-8.0 4.0-54.0
AB only (n=38)	115.8 \pm 33.8	103 93.0-138.0 3.6-300.0	4.0 \pm 4.3	3.0 2.0-4.0 1.0-21.0	3.2 \pm 2.2	3.0 2.0-3.0 1.0-17.0	553.3 \pm 948.6	354 200.0-560.0 3.6-5,954.0	12.6 \pm 17.3	5.5 3.0-11.0 2.0-77.0
All minor procedures (n=133)	117.3 \pm 39.9	100 90.0-140.0 3.6-300.0	3.3 \pm 3.3	2.0 2.0-3.0 1.0-21.0	5.9 \pm 43.2	2.0 2.0-3.0 1.0-749.0	458.1 \pm 962.4	270 180.0-420.0 3.6-8,544.0	17.0 \pm 74.3	5.0 3.0-8.0 2.0-749.0

Patient group/ surgical category	Dosage ($\mu\text{g}/\text{kg}$)		Number of doses		Interval between doses (h)		Cumulative dose ($\mu\text{g}/\text{kg}$)		Duration of treatment (h)	
	Mean $\pm\text{SD}$	Median IQR Range	Mean $\pm\text{SD}$	Median IQR Range	Mean $\pm\text{SD}$	Median IQR Range	Mean $\pm\text{SD}$	Median IQR Range	Mean $\pm\text{SD}$	Median IQR Range
<i>B. Major procedures (n=37)</i>										
No	90.0	90	11.1	11.0	4.7	3.0	1,594.0	979	52.4	48.0
AB/refractoriness (n=16)	± 28.9	90.0-90.0 28.0-180.0	± 8.4	3.0-20.0 1.0-24.0	± 4.3	2.0-6.0 1.0-30.0	$\pm 1,835.$	270.0-1,983.5 180.0-6,336.0	± 43.6	9.0-73.0 4.0-139.0
AB+refractoriness (n=0)	–	–	–	–	–	–	–	–	–	–
Refractoriness only (n=2)	122.3 ± 25.7	142 90.0-142.0 90.0-142.0	14.5 ± 4.9	14.5 11.0-18.0 11.0-18.0	3.2 ± 1.8	3.0 2.0-3.0 1.0-9.0	2,662.0 $\pm 2,364.$	2,662 990.0-4,334.0 990.0-4,334.0	43.5 ± 21.9	43.5 28.0-59.0 28.0-59.0
AB only (n=7)	82.9 ± 37.1	90 90.0-92.0 25.0-240.0	10.3 ± 10.3	2.0 2.0-21.0 2.0-22.0	10.2 ± 32.8	4.0 4.0-10.0 1.0-264.0	1,449.0 $\pm 1,658.$	480 180.0-2,711.0 180.0-4,424.0	107.0 ± 105.9	86.5 3.0-199.0 3.0-264.0
All major procedures (n=25)	91.5 ± 32.7	90 90.0-92.0 25.0-240.0	11.1 ± 8.5	11.0 3.0-21.0 1.0-24.0	5.9 ± 17.0	3.0 2.0-6.0 1.0-264.0	1,638.8 $\pm 1,767.$	990 270.0-2,167.0 180.0-6,336.0	66.5 ± 67.3	48.5 9.0-97.0 3.0-264.0

AB: platelet antibodies; IQR: interquartile range; refractoriness: platelet refractoriness.

Supplementary Table 2. Cumulative rFVIIa usage in rFVIIa, rFVIIa+AF and rFVIIa+P±AF treatments.

Patient group/surgical category	Statistics	rFVIIa	rFVIIa+AF	rFVIIa+P±AF
<i>A. Minor procedures</i>				
No AB/refractoriness	N procedures	24	16	4
	Mean±SD	206.6±71.2	718.4±2,089.9	268.4±181.1
	Median	180	180	180
AB+refractoriness	N procedures	18	19	3
	Mean±SD	432.9±276.9	336.9±191.5	252.7±50.3
	Median	417.5	300	270
Refractoriness only	N procedures	4	5	2
	Mean±SD	203.3±131.5	325.4±128.6	2,488.5±3,010.2
	Median	195	300	2,488.5
AB only	N procedures	10	24	4
	Mean±SD	268.2±145.8	742.8±1,155.0	129.2±88.3
	Median	253	420	156.5
All minor procedures	N procedures	56	64	13
	Mean±SD	290.1±201.5	583.6±1,255.5	563.5±1,224.5
	Median	250.5	300	196
<i>B. Major procedures</i>				
No AB/refractoriness	N procedures	4	3	9
	Mean±SD	3,354±2,875.9	780±883.3	1,083.1±963.4
	Median	3,360	270	463
AB+refractoriness	N procedures	–	–	–
	Mean±SD	–	–	–
	Median	–	–	–
Refractoriness only	N procedures	–	–	2
	Mean±SD	–	–	2,662.0±2,364.6
	Median	–	–	2,662
AB only	N procedures	2	4	1
	Mean±SD	330±212.1	2,325.8±1,757.8	180
	Median	330	2,349.5	180
All major procedures	N procedures	6	7	12

Mean±SD	2,346±2,722.2	1,663.3±1,577.2	1,271.0 ±1,293.1
Median	987.5	1800	726.5

Mean and median doses are in µg/kg.

AB: platelet antibodies; AF: antifibrinolytic agent(s); P: platelets; refractoriness: platelet refractoriness; rFVIIa: recombinant factor

VIIa; SD: standard deviation.