

The status of care for persons with von Willebrand disease registered within CNHP registry Annual Report 2019

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on behalf of

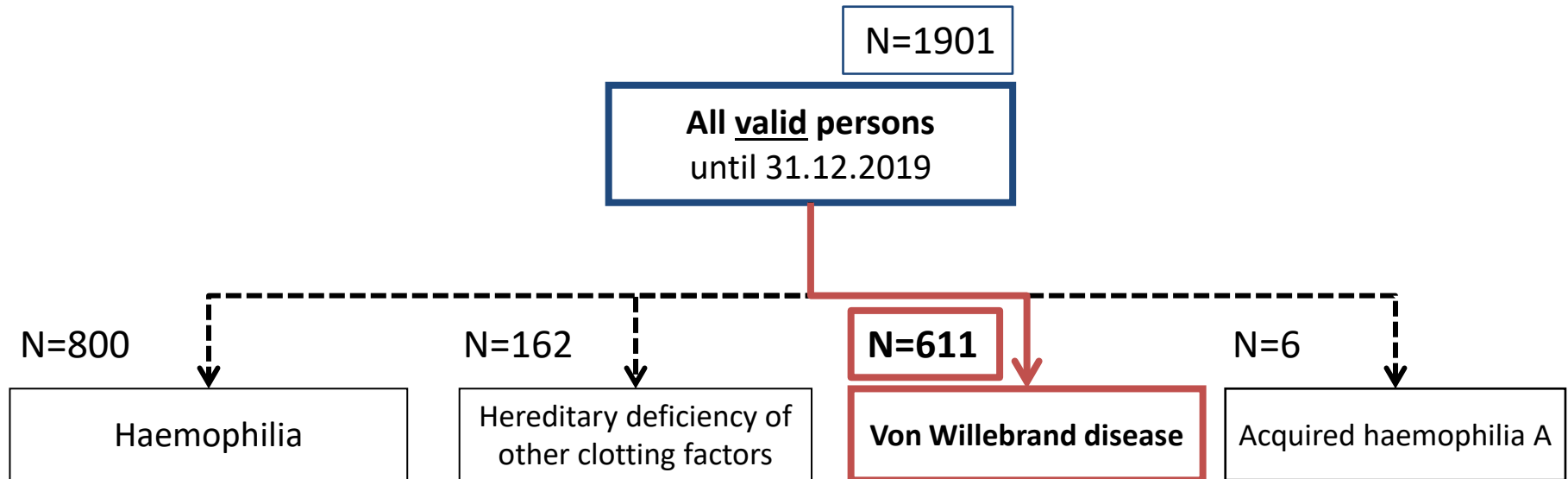
Centres contributing to database
of the CNHP (Czech National Haemophilia Programme)

Export date: May 11, 2020

Persons with Von Willebrand disease



Sample size



Cca 1000 symptomatic vWDs should be in CZ
611 of them are already in CNHP registry

Number of patients in participating centres

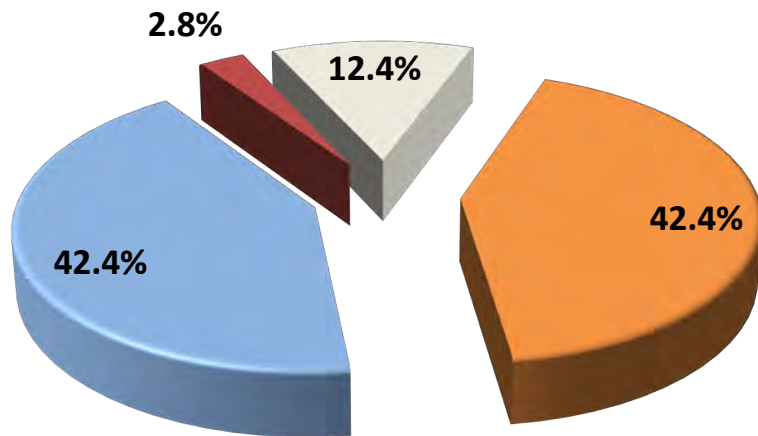
N=611





Paediatric centres	Valid patients	
	N	%
Prague – Dpt. of Pediatric Haematology and Oncology, CUH Motol	30	4.9
Pilsen – Pediatric Dpt., UH Pilsen	24	3.9
Ostrava – Dpt. of Pediatric Medicine, UH Ostrava	21	3.4
Hradec Králové – Dpt. of Pediatric Medicine, UH HK	20	3.3
Brno – Dpt. of Pediatric Haematology, CUH Brno	17	2.8
Ústí n.L. – Pediatric Dpt. – Haematology, Masaryk Hospital	14	2.3
Olomouc – Dpt. of Pediatric Medicine, UH Olomouc	1	0.2

Adult centres	Valid patients	
	N	%
Brno – Dpt. Of Clin Hematol, UH Brno	272	44.5
Pilsen – Dpt. of Biochemistry and Hematology, UH Pilsen – UKBH	77	12.6
Ostrava – Blood centre, UH Ostrava	68	11.1
Liberec – Dpt. Of Clin Hematol, Hospital Liberec	25	4.1
Olomouc – Haemato-Oncology Dpt., UH Olomouc	23	3.8
Ústí n.L. – Dpt. Of Clin Hematol, Masaryk Hospital	16	2.6
Hradec Králové – IV. Internal and Hematology Dpt., UH HK	2	0.3
České Budějovice – Dpt. Of Clin Hematol, Hospital CB	1	0.2

Type of Von Willebrand's disease

N=611



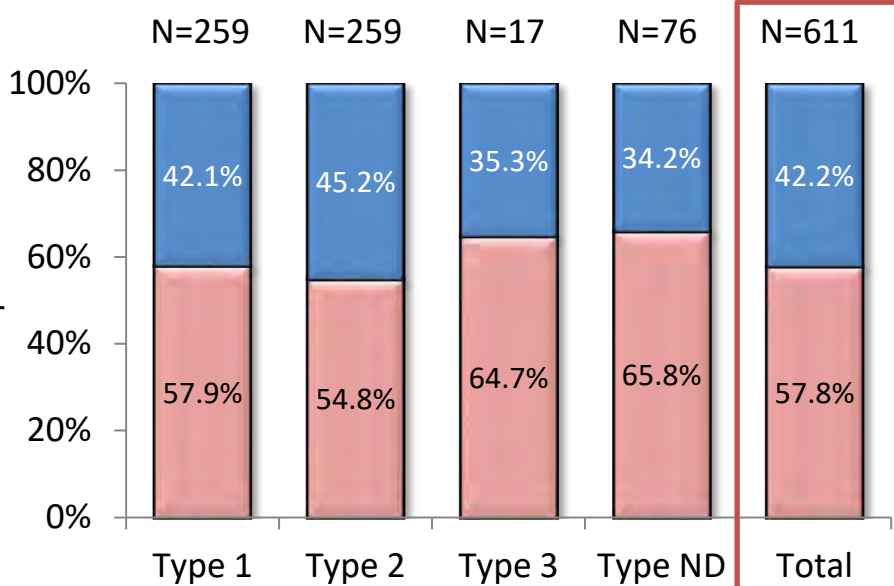
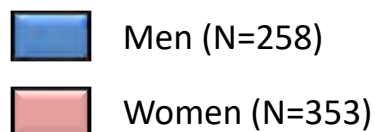
		Patients	
Type of disease		N	%
	Type 1*	259	42.4
	Type 2	259	42.4
	Type 2A	110	18.0
	Type 2B	20	3.3
	Type 2M	41	6.7
	Type 2N	22	3.6
	Type 2 (not specified)	66	10.8
	Type 3	17	2.8
	Type not determined / unfilled	76	12.4
Total		611	100.0

* of them 6 patients with type 1/2N

Sex and current age of patients

N=611

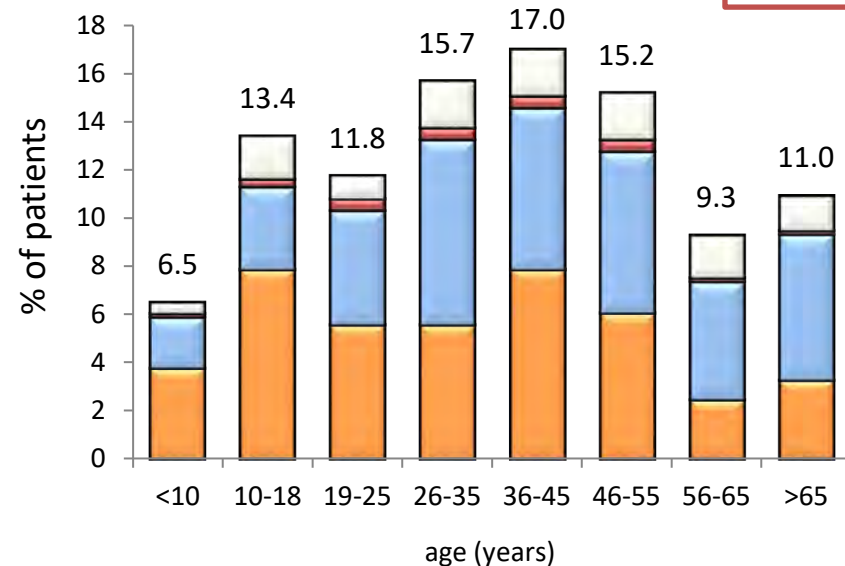
Sex



Type ND = not determined or unfilled

Current age*

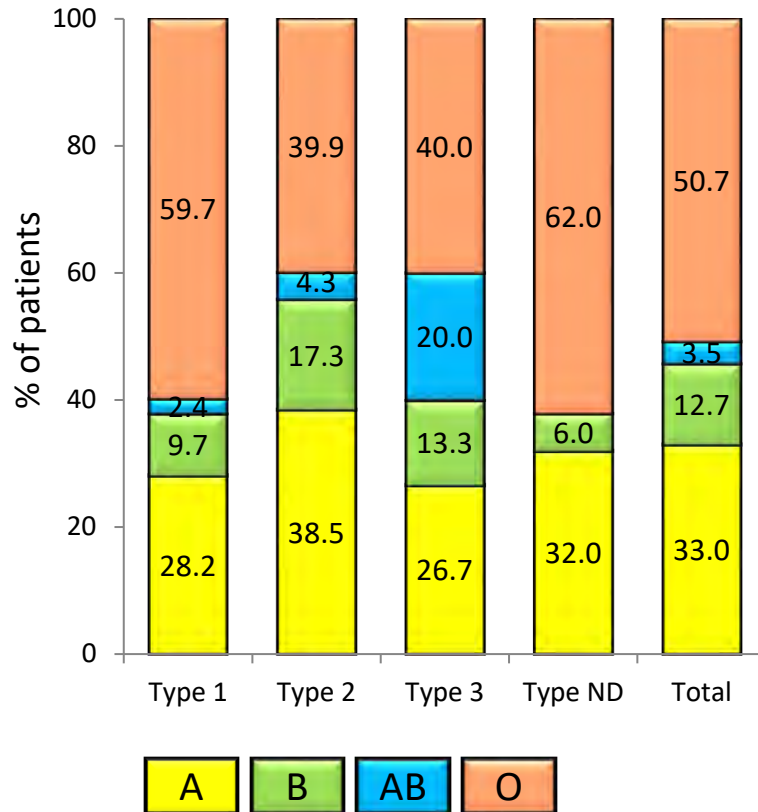
	Type 1	Type 2	Type 3	Type ND	Total
N	259	259	17	76	611
Mean	34.1	41.6	34.5	40.9	38.1
Median	33	41	33	40	37
min - max	2 - 91	2 - 86	3 - 68	5 - 82	2 - 91



* age reached in year 2019

Blood group

N=611

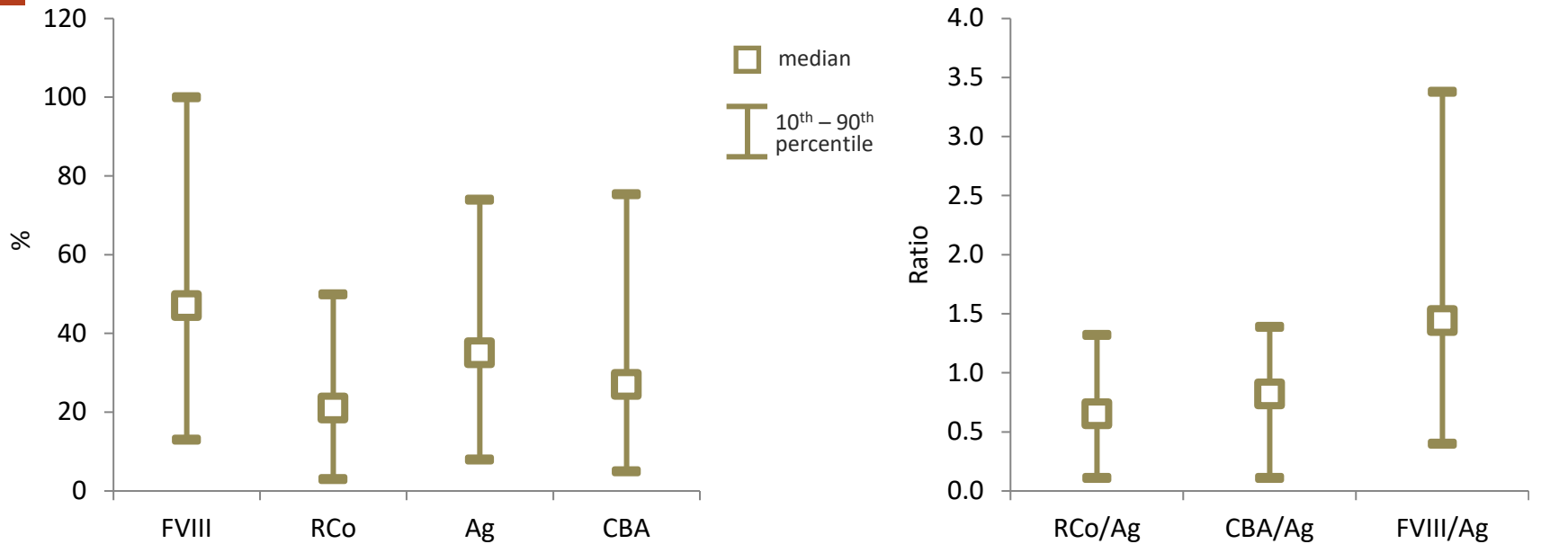


%¹ - % of total N
 %² - % of valid N

Blood group	A	B	AB	O	ND	Valid N	Total N
Type 1	<i>N</i>	58	20	5	123	53	206
	% ¹	22.4	7.7	1.9	47.5	20.5	100.0
	% ²	28.2	9.7	2.4	59.7		100.0
Type 2	<i>N</i>	80	36	9	83	51	208
	% ¹	30.9	13.9	3.5	32.0	19.7	100.0
	% ²	38.5	17.3	4.3	39.9		100.0
Type 3	<i>N</i>	4	2	3	6	2	15
	% ¹	23.5	11.8	17.6	35.3	11.8	100.0
	% ²	26.7	13.3	20.0	40.0		100.0
Type ND	<i>N</i>	16	3	0	31	26	50
	% ¹	21.1	3.9	0.0	40.8	34.2	100.0
	% ²	32.0	6.0	0.0	62.0		100.0
Total	<i>N</i>	158	61	17	243	132	479
	% ¹	25.9	10.0	2.8	39.8	21.6	100.0
	% ²	33.0	12.7	3.5	50.7		100.0

Factor levels

N=611

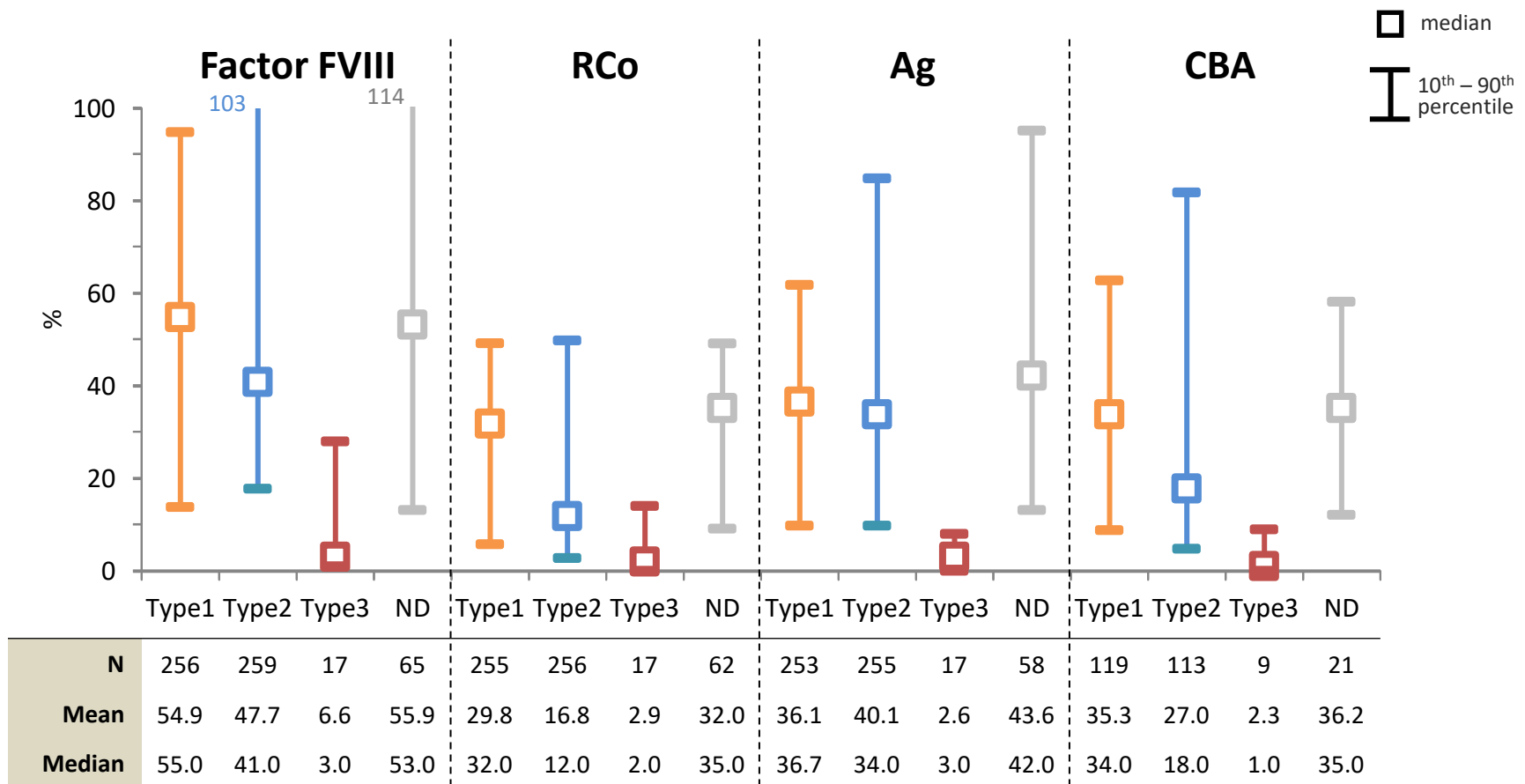


Factor FVIII*	RCo	Ag	CBA		RCo/Ag	CBA/Ag	FVIII/Ag
597	590	583	262	N	575	261	579
50.5	23.6	37.6	30.6	Mean	0.8	0.8	1.9
47 (1–232)	21 (0–94)	35 (0–261)	27 (0–127)	Median (min – max)	0.65 (0–38)	0.82 (0–3)	1.44 (0.08–106)

* Factor FVIII was assessed by coagulation method in 549 patients, by chromogenic method in 17 patients and method is missing in 44 patients

Factor levels according to type of VWD, part 1

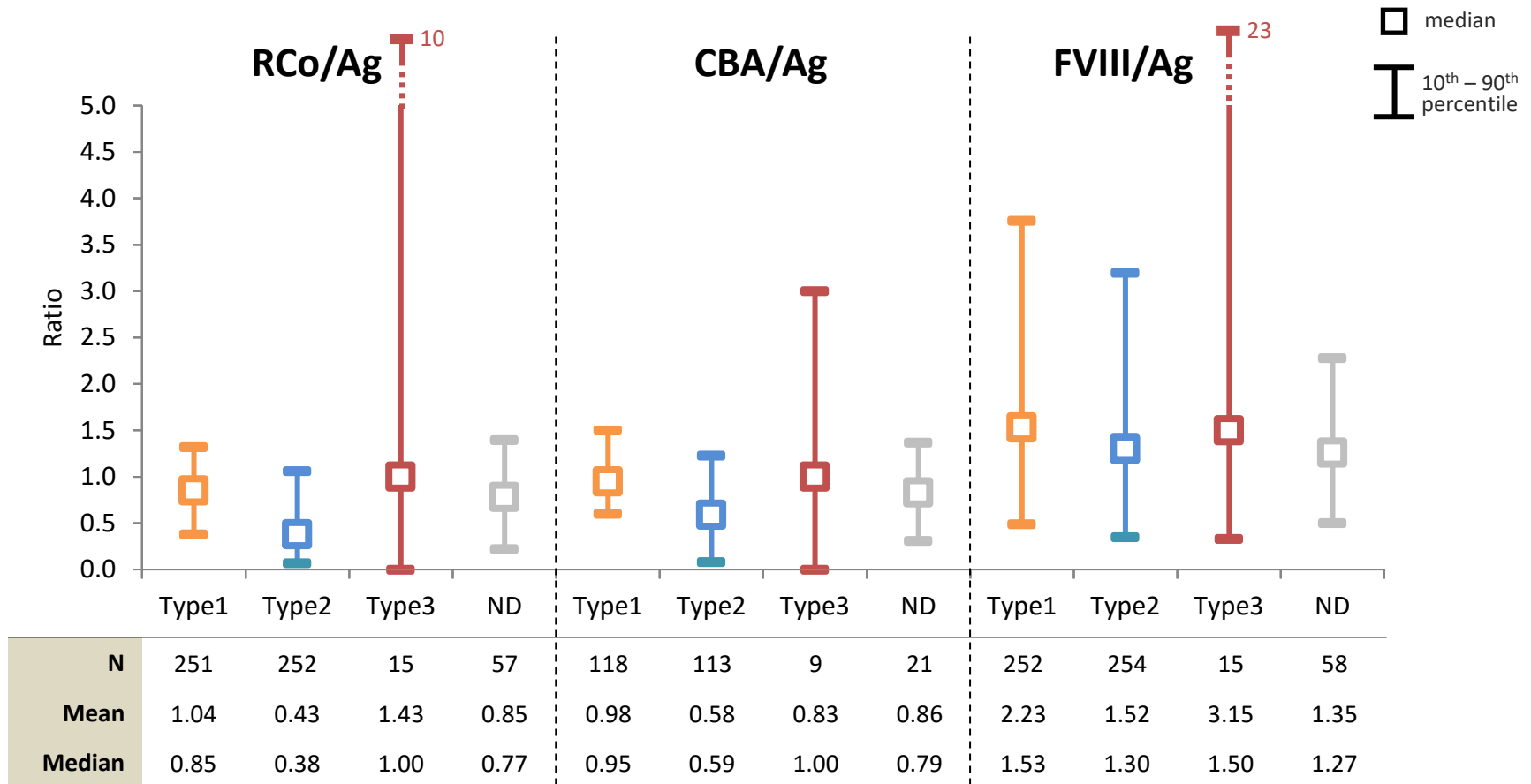
N=611



Type ND = not determined or unfilled

Factor levels according to type of VWD, part 2

N=611

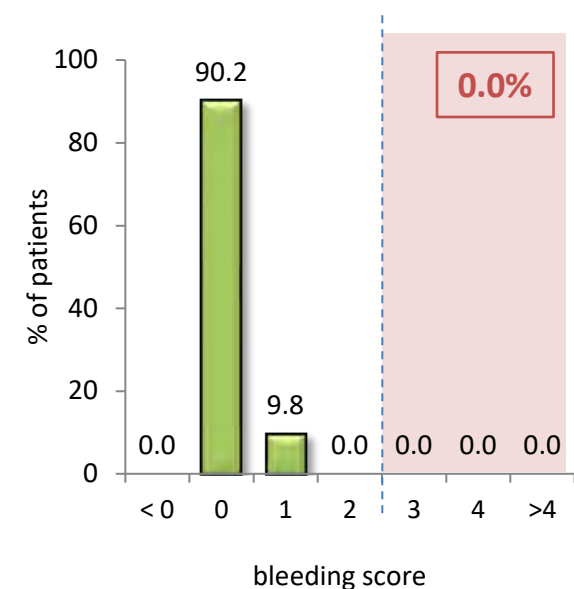
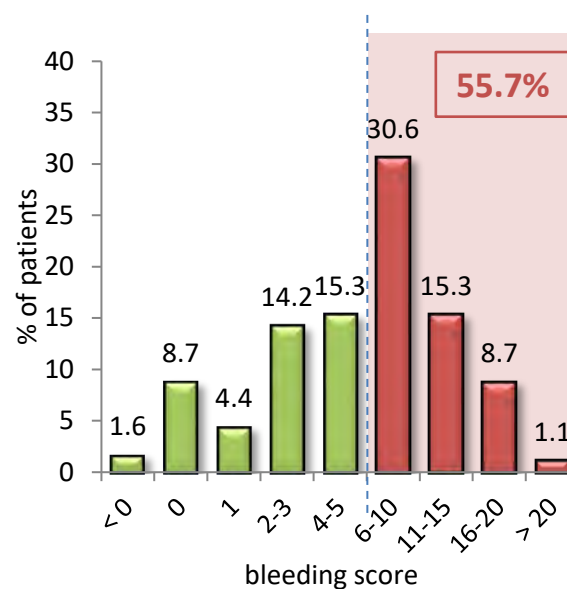
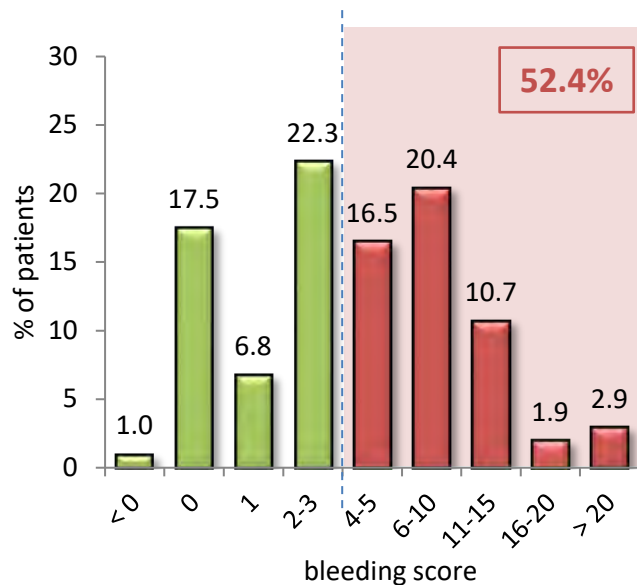


Type ND = not determined or unfilled

Bleeding score¹ according to sex and age

N=368²

	Adult men	Adult women	Children
N	103	183	82
Mean	5.2	7.2	0.1
Median (min - max)	4 (-1 - 25)	6 (-2 - 24)	0 (0 - 1)



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

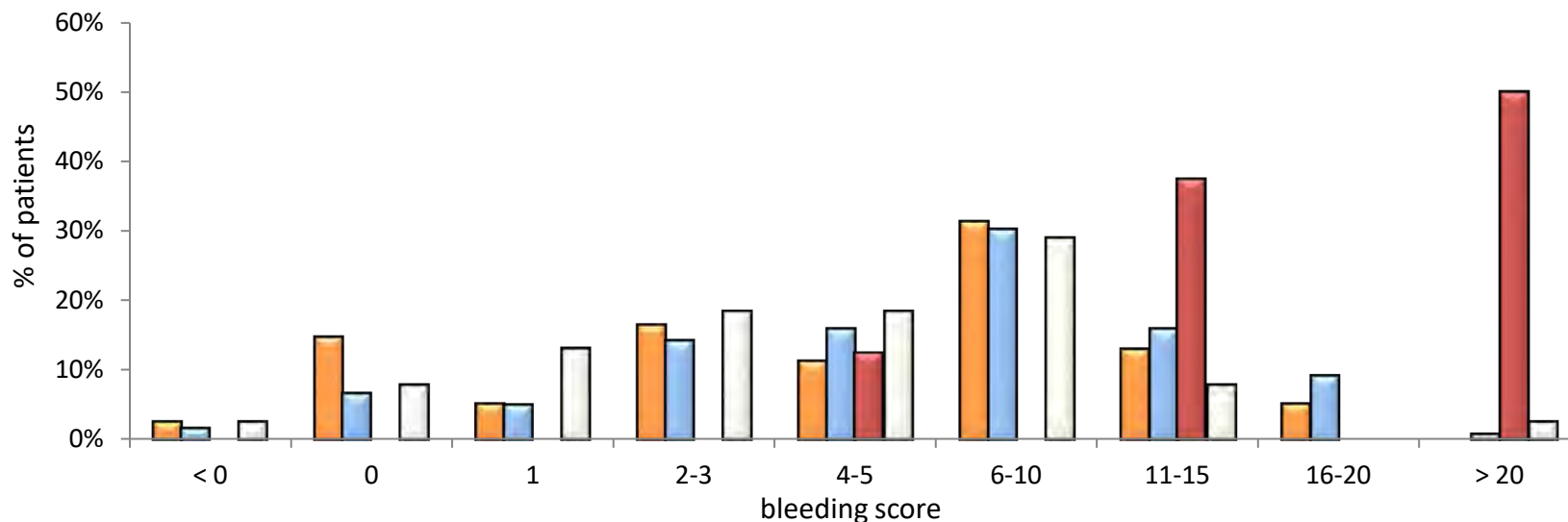
² Missing information on bleeding score in 243 patients.

Bleeding score¹ in adults according to type of disease

N=286²

Bleeding score

	Type 1	Type 2	Type 3	Type ND	Total
N total	188	225	14	62	489
N valid	129	129	8	20	286
Mean	5.6	6.8	17.1	5.8	6.5
Median	5.0	6.0	17.5	5.0	5.0
min - max	-2-20	-1-22	5-25	0-14	-2-25



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

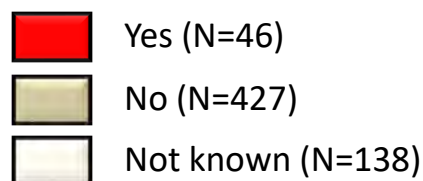
² Adult patients with non-missing information on bleeding score.

Type ND = not determined or unfilled

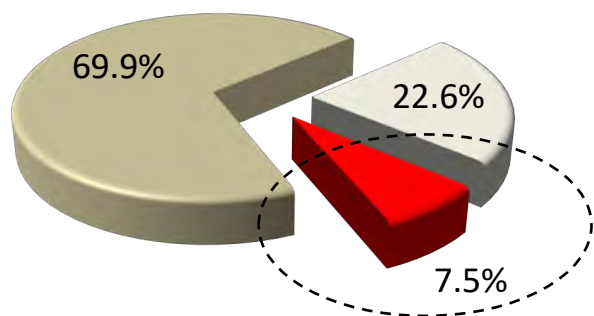
Concomitant diseases

N=611

Experienced hepatitis



None of the patients is HIV positive.



*28x hepatitis A
9x hepatitis B
5x hepatitis A+B
2x hepatitis C
1x hepatitis A+C
1x hepatitis A+B+C*

Annual bleeding rate (ABR) and location of bleeds treated with factor concentrate

N=611

	Type 1	Type 2	Type 3	Type ND	Total
ABR					
N valid	259	259	17	76	611
Mean	0.06	0.22	2.12	0.04	0.18
Median	0	0	0	0	0
min – max	0 - 4	0 - 10	0 - 14	0 - 3	0 - 14
SUM	15	57	36	3	111
<hr/>					
Location of bleeds					
N_p/N_B^2					
Joints	0/-	3/3	4/8	0/-	7/11
Epistaxes	4/9	12/20	2/10	1/1	19/40
Urogenital tract	0/-	2/2	1/2	0/-	3/4
GIT	0/-	2/2	1/1	0/-	3/3
Subcutaneous	0/-	1/1	1/2	0/-	2/3
Muscles	0/-	2/2	2/5	0/-	4/7
CNS	0/-	0/-	0/-	0/-	0/-
Metrorrhagia	0/-	0/-	1/4	0/-	1/4
Other	2/2	6/6	3/4	1/2	12/14
SUM	6/11	28/36	15/36	2/3	51/86

² N_p = number of patients with bleed;

Type ND = not determined or unfilled

N_B = total number of bleeds treated with substitution of VWF

Consumption of VWF concentrates in year 2019

N=611

	Number of patients treated with substitution of VWF total (type1/type2/type3/typeND)	Total annual consumption (IU)	Average annual consumption per treated patient (IU)
Fanhdi	8 (3/5/0/0)	68 000	8 500.0
<i>of them on prophylaxis*</i>	<i>1 (0/1/0/0)</i>	<i>39 000</i>	<i>39 000.0</i>
Haemate P	72 (16/42/8/6)	2 023 000	28 097.2
<i>of them on prophylaxis*</i>	<i>3 (0/1/2/0)</i>	<i>541 500</i>	<i>180 500.0</i>
Immunate	2 (2/0/0/0)	2 500	1 250.0
<i>of them on prophylaxis*</i>	<i>0 (0/0/0/0)</i>	<i>0</i>	<i>0.0</i>
Wilate	21 (9/9/3/0)	220 000	10 476.2
<i>of them on prophylaxis*</i>	<i>1 (0/0/1/0)</i>	<i>91 000</i>	<i>91 000.0</i>
Willfact	1 (0/0/1/0)	2 000	2 000.0
<i>of them on prophylaxis*</i>	<i>1 (0/0/1/0)</i>	<i>2 000</i>	<i>2 000.0</i>
Total	104 (30/56/12/6)	2 313 000	21 459.3
<i>of them on prophylaxis*</i>	<i>6 (0/2/4/0)</i>	<i>673 500</i>	<i>112 250.0</i>
Total - type 1	30	251 502	<u>8 193.6</u>
Total - type 2	56	1 087 999	<u>18 476.3</u>
Total - type 3	12	877 000	<u>73 083.3</u>
Total - type ND	6	96 500	<u>16 083.3</u>

* permanent prophylaxis

Type ND = not determined or unfilled

Consumption of other drugs in year 2019

N=611

	Number of patients treated with other drugs total (type1/type2/type3/typeND)	Total annual consumption	Average annual consumption per <u>treated</u> patient
DDAVP (µg)	4 (1/3/0/0)	2 102	525.4
<i>of them on prophylaxis*</i>	<i>0 (0/0/0/0)</i>	<i>0</i>	<i>0.0</i>

* *permanent prophylaxis*

Type ND = not determined or unfilled

Patients on permanent prophylaxis in detail

N=4

No	Centre	Age group	Type of VWD	Type of prophylaxis in 2019	Treatment in 2019	No of applications per week	Total consumption (IU)	ABR in 2019
1	Brno	Adult	3	Permanent	Haemate P	2	235 000	3
2	Brno	Adult	3	Permanent	Haemate P	2	294 000	4
3	Olomouc	Adult	3	Permanent	Willfact, Wilate	N/A	93 000	0
4	Plzeň	Adult	2	Permanent	Fanhdi, Haemate P, DDAVP	1.5	51 500 (w/o DDAVP)	0