

The status of care for persons with von Willebrand disease registered within CNHP registry Annual Report 2021

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on behalf of

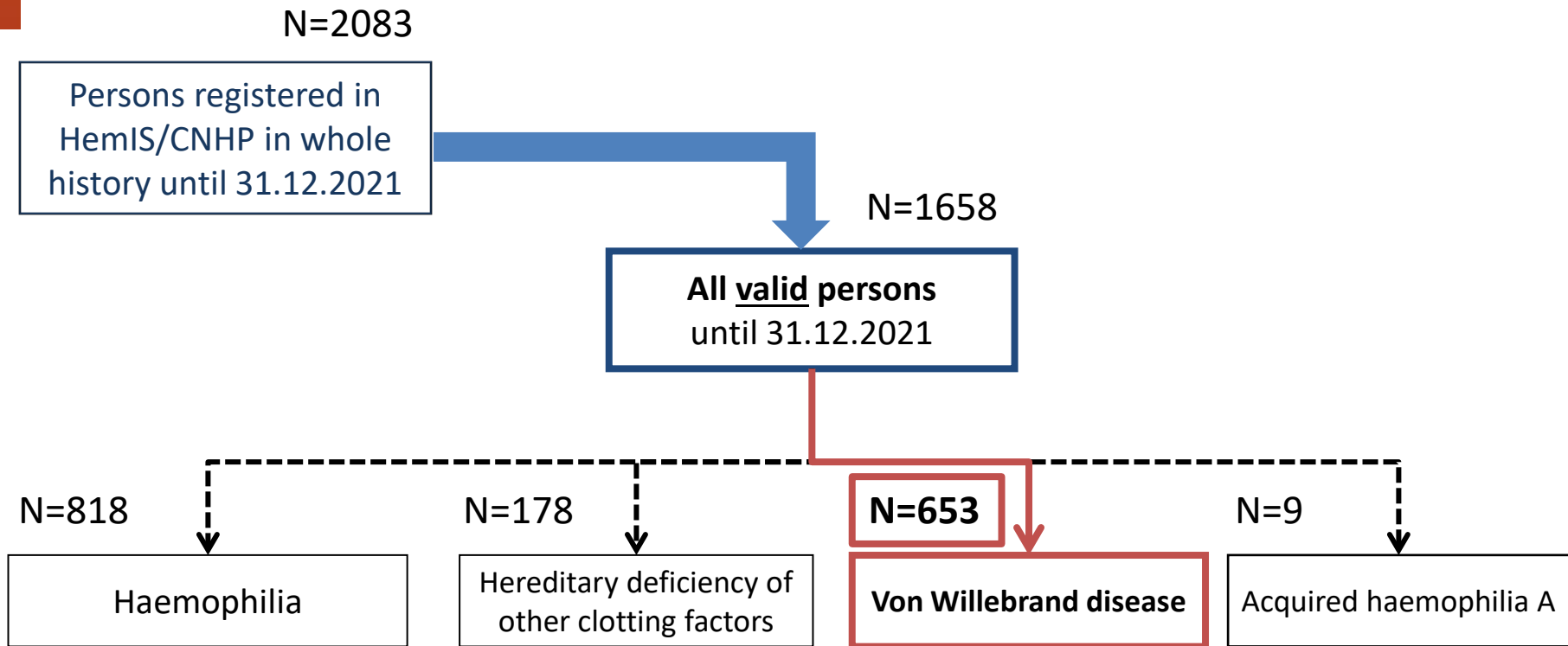
Centres contributing to database
of the CNHP (Czech National Haemophilia Programme)

Export date: April 01, 2022

Persons with Von Willebrand disease



Sample size



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

Number of patients in participating centres

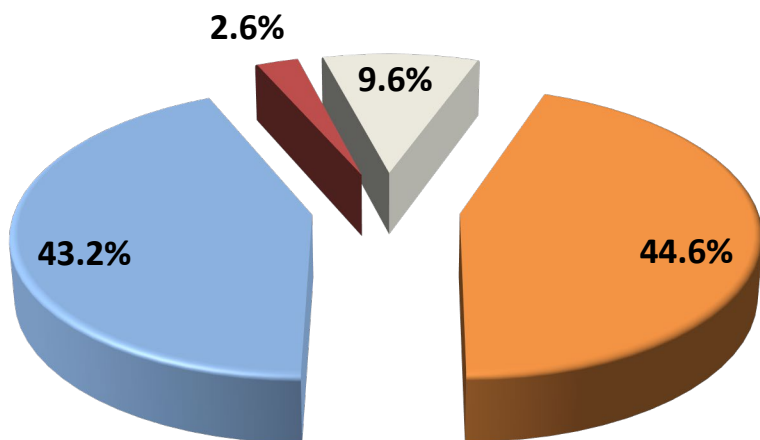
N=653





Paediatric centres	Valid patients	
	N	%
Prague – Dpt. of Pediatric Haematology and Oncology, CUH Motol	29	4.4
Ostrava – Dpt. of Pediatric Medicine, UH Ostrava	24	3.7
Pilsen – Pediatric Dpt., UH Pilsen	23	3.5
Hradec Králové – Dpt. of Pediatric Medicine, UH HK	21	3.2
Brno – Dpt. of Pediatric Haematology, CUH Brno	21	3.2
Ústí n.L. – Pediatric Dpt. – Haematology, Masaryk Hospital	11	1.7
Olomouc – Dpt. of Pediatric Medicine, UH Olomouc	10	1.5

Adult centres	Valid patients	
	N	%
Brno – Dpt. Of Clin Hematol, UH Brno	287	44.0
Pilsen – Dpt. of Biochemistry and Hematology, UH Pilsen – UKBH	79	12.1
Ostrava – Blood centre, UH Ostrava	72	11.0
Liberec – Dpt. Of Clin Hematol, Hospital Liberec	32	4.9
Olomouc – Haemato-Oncology Dpt., UH Olomouc	23	3.5
Ústí n.L. – Dpt. Of Clin Hematol, Masaryk Hospital	18	2.8
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=653



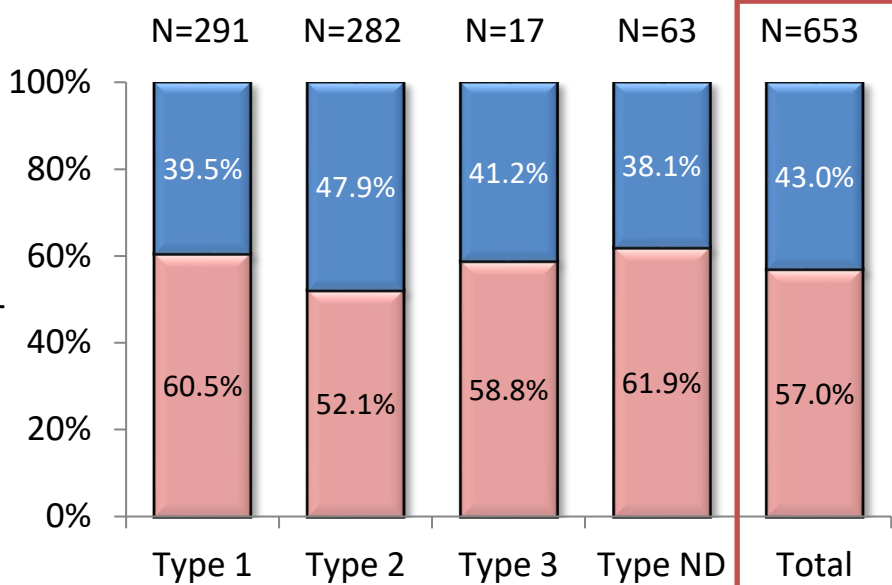
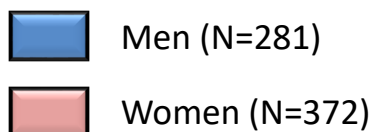
		Patients	
Type of disease		N	%
	Type 1*	291	44.6
	Type 2	282	43.2
	Type 2A	122	18.7
	Type 2B	18	2.8
	Type 2M	46	7.0
	Type 2N	25	3.8
	Type 2 (not specified)	71	10.9
	Type 3	17	2.6
	Type not determined / unfilled	63	9.6
Total		653	100.0

* of them 6 patients with type 1/2N

Sex and current age of patients

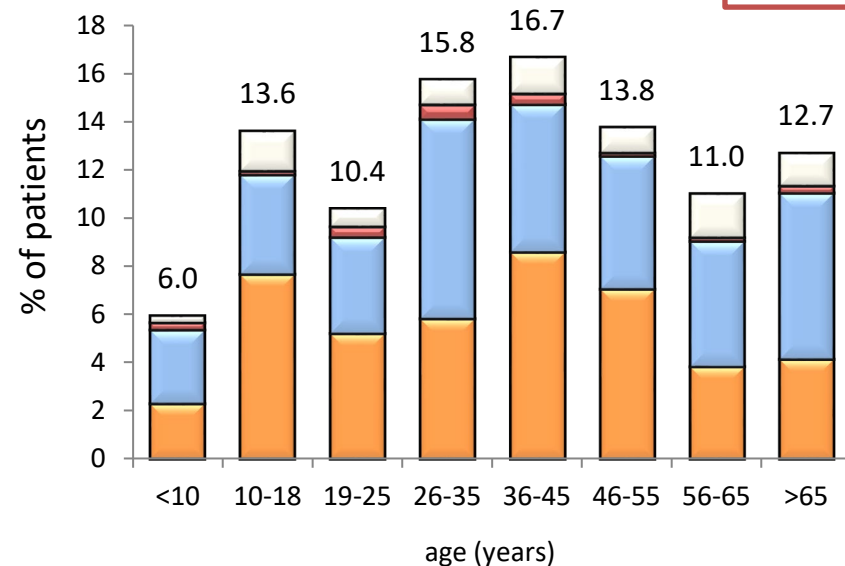
N=653

Sex



Current age*

	Type 1	Type 2	Type 3	Type ND	Total
N	291	282	17	63	653
Mean	37.1	40.9	33.8	42.3	39.2
Median	37	40	34	42	38
min - max	1-93	0-88	2-70	0-84	0-93

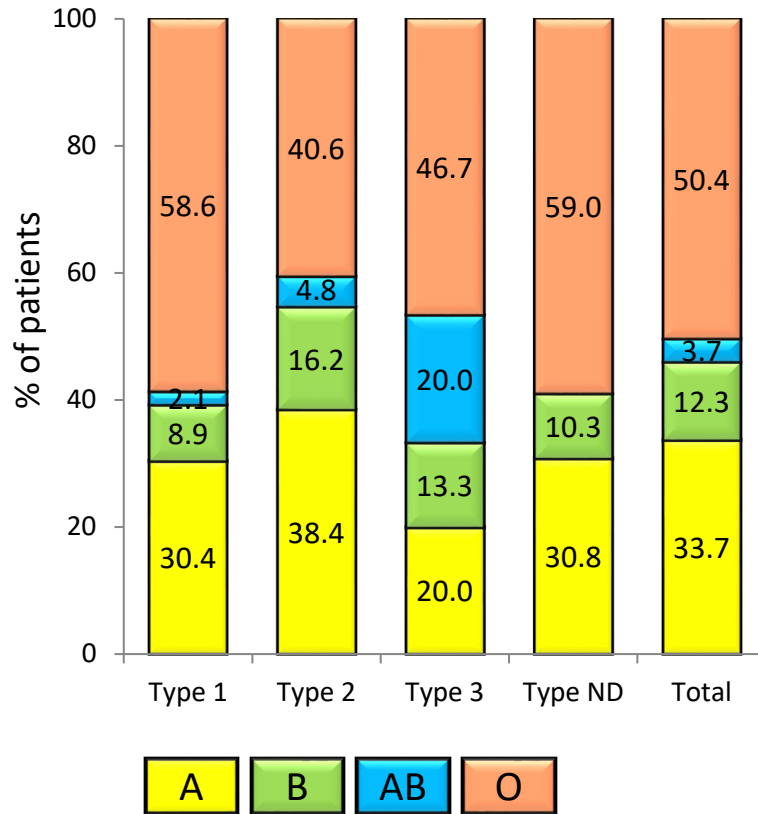


Type ND = not determined or unfilled

* age reached in year 2021

Blood group

N=653

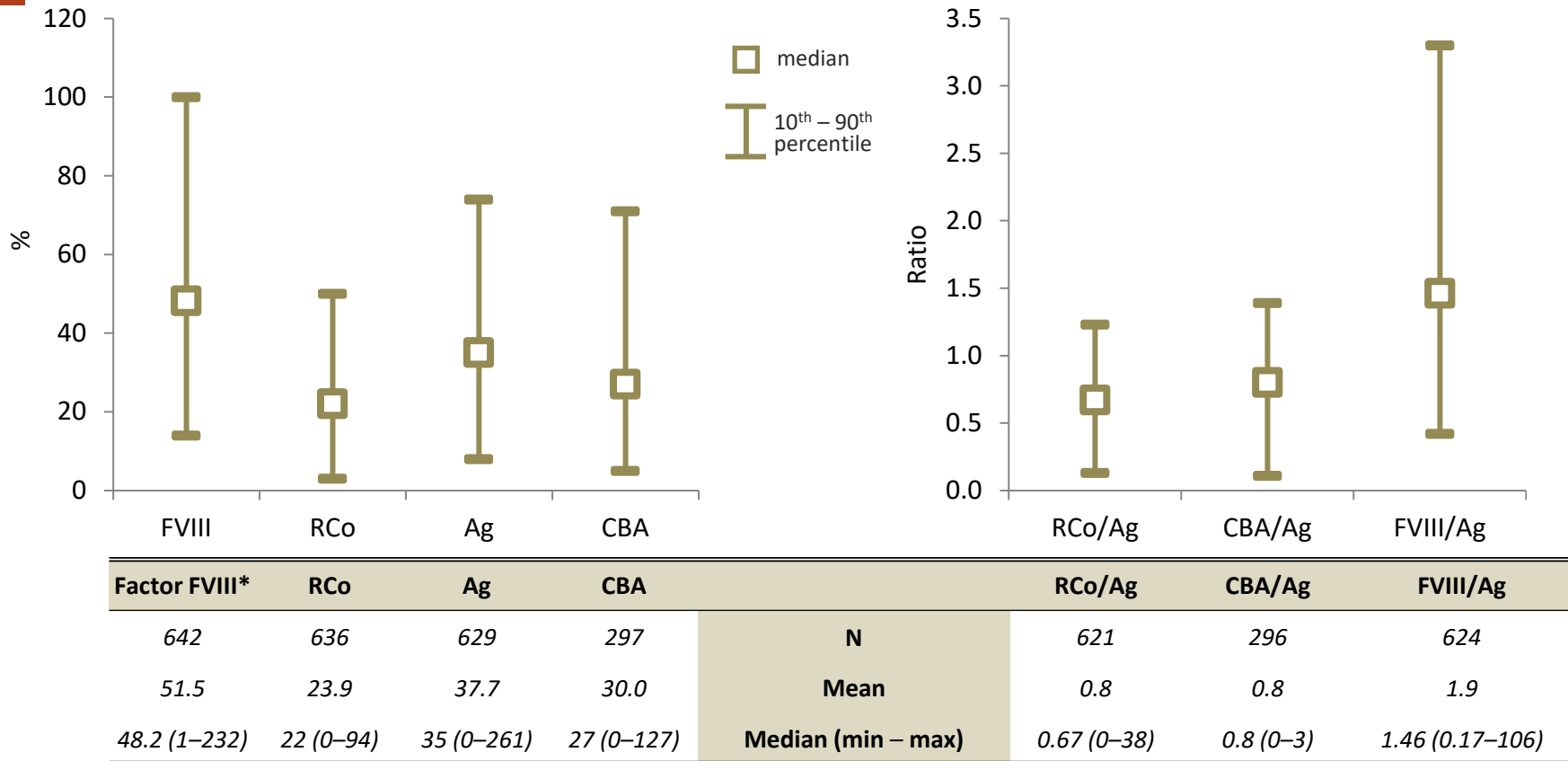


%¹ - % of total N
 %² - % of valid N

Blood group	A	B	AB	O	ND	Valid N	Total N
Type 1	<i>N</i>	72	21	5	139	54	237
	% ¹	24.7	7.2	1.7	47.8	18.6	100.0
	% ²	30.4	8.9	2.1	58.6	100.0	
Type 2	<i>N</i>	88	37	11	93	53	229
	% ¹	31.2	13.1	3.9	33.0	18.8	100.0
	% ²	38.4	16.2	4.8	40.6	100.0	
Type 3	<i>N</i>	3	2	3	7	2	15
	% ¹	17.6	11.8	17.6	41.2	11.8	100.0
	% ²	20.0	13.3	20.0	46.7	100.0	
Type ND	<i>N</i>	12	4	0	23	24	39
	% ¹	19.0	6.3	0.0	36.5	38.1	100.0
	% ²	30.8	10.3	0.0	59.0	100.0	
Total	<i>N</i>	175	64	19	262	133	653
	% ¹	26.8	9.8	2.9	40.1	20.4	100.0
	% ²	33.7	12.3	3.7	50.4	100.0	

Factor levels

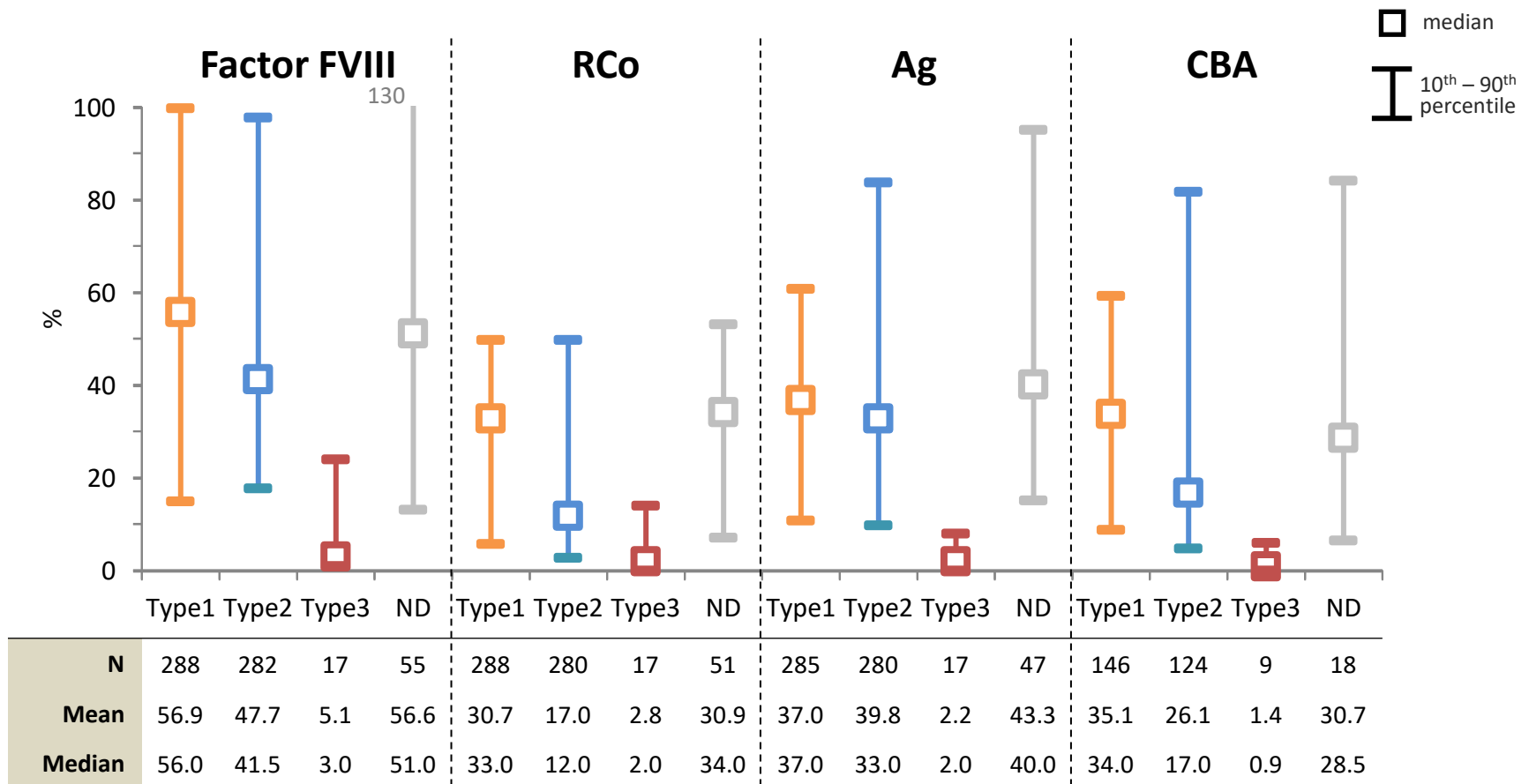
N=653



* Factor FVIII was assessed by coagulation method in 595 patients, by chromogenic method in 18 patients and method is missing in 29 patients.

Factor levels according to type of VWD, part 1

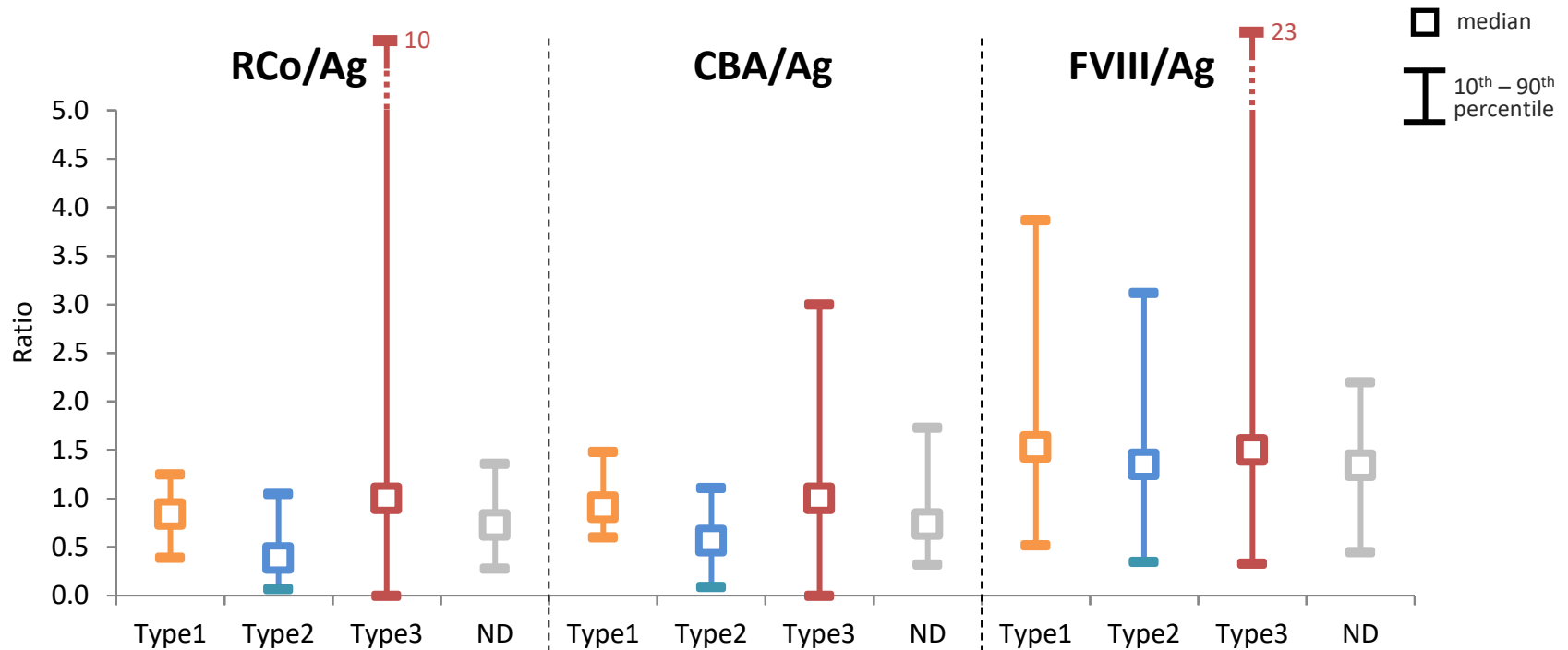
N=653



Type ND = not determined or unfilled

Factor levels according to type of VWD, part 2

N=653



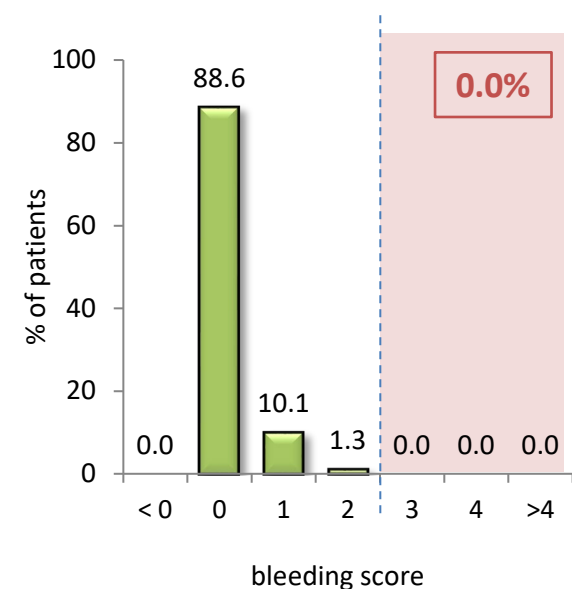
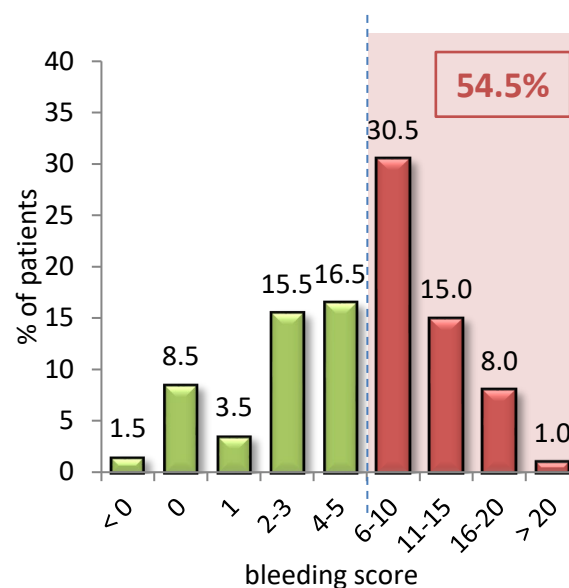
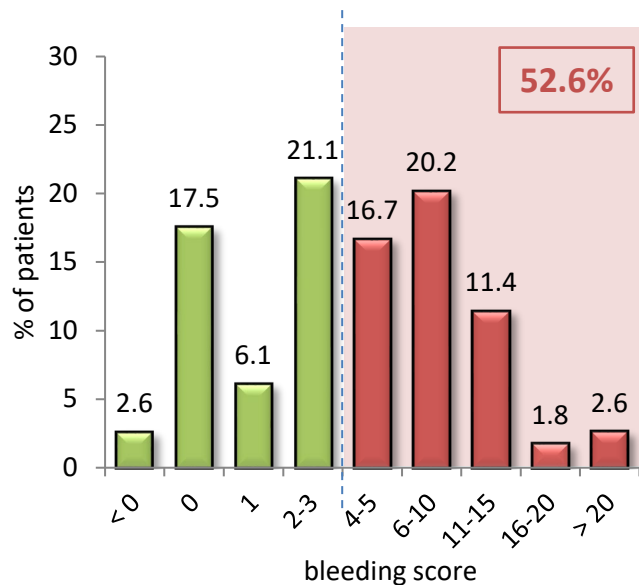
	Type1	Type2	Type3	ND	Type1	Type2	Type3	ND	Type1	Type2	Type3	ND
N	284	276	15	46	145	124	9	18	284	278	15	47
Mean	1.02	0.44	1.53	0.81	0.96	0.57	0.89	0.80	2.19	1.52	3.59	1.36
Median	0.84	0.39	1.00	0.73	0.91	0.57	1.00	0.74	1.53	1.35	1.50	1.34

Type ND = not determined or unfilled

Bleeding score¹ according to sex and age

N=393²

	Adult men	Adult women	Children
N	114	200	79
Mean	5.2	7.0	0.1
Median (min - max)	4 (-2 - 25)	6 (-2 - 24)	0 (0 - 2)



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

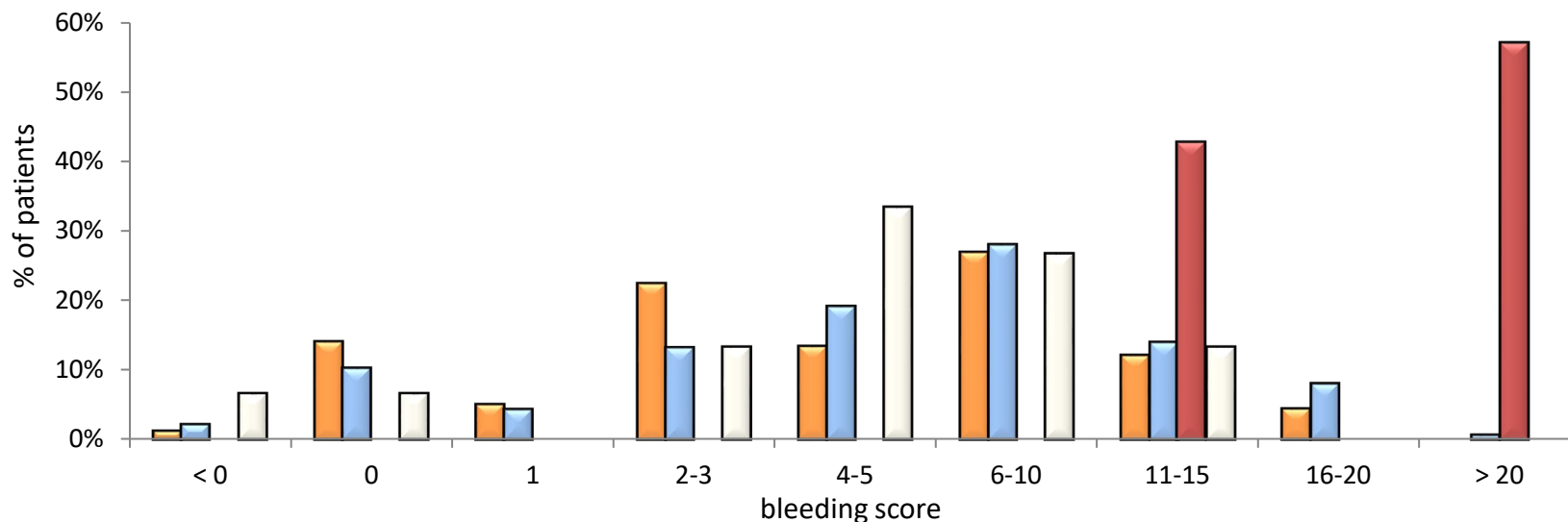
² Missing information on bleeding score in 260 patients.

Bleeding score¹ in adults according to type of disease

N=314²

Bleeding score

	Type 1	Type 2	Type 3	Type ND	Total
N total	226	235	14	50	525
N valid	156	136	7	15	314
Mean	5.5	6.7	18.9	5.7	6.4
Median	5.0	6.0	21.0	5.0	5.0
min - max	-2-20	-1-22	13-25	-2-15	-2-25



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

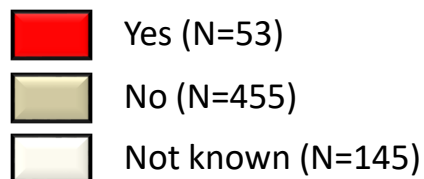
² Adult patients with non-missing information on bleeding score.

Type ND = not determined or unfilled

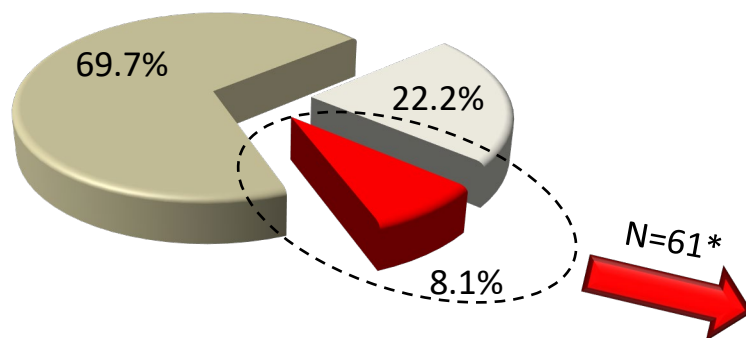
Concomitant diseases

N=653

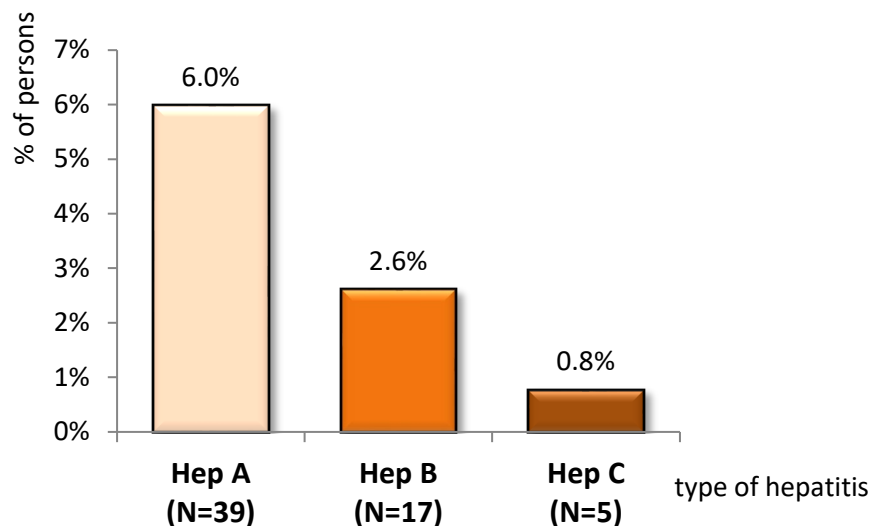
Experienced hepatitis



None of the patients is HIV positive.



*Total of 61 cases of hepatitis in 53 persons. One person may have more types of hepatitis recorded.



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

	Type 1	Type 2	Type 3	Type ND	Total	N=653
ABR						
N valid	291	282	17	63	653	
Mean	0.03	0.10	2.53	0.02	0.12	
Median	0	0	0	0	0	
min-max	0-3	0-9	0-20	0-1	0-20	
SUM	10	27	43	1	81	
<hr/>						
Location of bleeds						
N_p/N_B^2						
Joints	1/1	0/-	2/4	0/-	3/5	
Epistaxes	1/1	8/11	3/23	0/-	12/35	
Urogenital tract	0/-	1/1	0/-	0/-	1/1	
GIT	0/-	1/1	1/10	0/-	2/11	
Subcutaneous	0/-	3/3	0/-	0/-	3/3	
Muscles	1/1	0/-	4/4	0/-	5/5	
CNS	0/-	0/-	0/-	0/-	0/-	
Metrorrhagia	1/3	0/-	0/-	0/-	1/3	
Other	1/1	6/6	2/2	0/-	9/9	
SUM	5/7	19/22	12/43	0/-	36/72	

² N_p = number of patients with bleed;

N_B = total number of bleeds treated with substitution of VWF

Type ND = not determined or unfilled

Consumption of VWF concentrates in year 2021

N=653

	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)	134 500	16 812.5
<i>of them on prophylaxis*</i>	<i>1 (0/1/0/0)</i>	<i>44 000</i>	<i>44 000.0</i>
Haemate P	66 (20/37/7/2)	1 672 500	25 340.9
<i>of them on prophylaxis*</i>	<i>3 (0/0/3/0)</i>	<i>794 000</i>	<i>264 666.7</i>
Wilate	20 (10/6/2/2)	354 000	17 700.0
<i>of them on prophylaxis*</i>	<i>2 (0/0/2/0)</i>	<i>212 000</i>	<i>106 000.0</i>
Total	94 (33/48/9/4)	2 161 000	22 497.9
<i>of them on prophylaxis*</i>	<i>6 (0/1/5/0)</i>	<i>1 050 000</i>	<i>175 000.0</i>
Total - type 1	33	350 026	<u>10 000.7</u>
Total - type 2	48	590 475	<u>12 302.1</u>
Total - type 3	9	1 133 000	<u>125 888.9</u>
Total - type ND	4	87 500	<u>21 875.0</u>

* permanent prophylaxis

Type ND = not determined or unfilled

Consumption of other drugs in year 2021

N=653

	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)	2 (2/0/0/0)	26	12.8
<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=6

No	Centre	Age group	Type of VWD	Type of prophylaxis in 2021	Treatment in 2021	No of applications per week	Total consumption (IU)	ABR in 2021
1	Praha	Child	3	Permanent	Wilate	3	160 000	20
2	Ostrava	Child	3	Permanent	Wilate	2	52 000	1
3	Plzeň	Adult	2	Permanent	Fanhdi + Haemate P	1	52 500	0
4	Brno	Adult	3	Permanent	Haemate P	2.3	312 000	2
5	Brno	Adult	3	Permanent	Haemate P	3.3	300 000	3
6	Brno	Adult	3	Permanent	Haemate P	2	182 000	0