

# The status of care for persons with von Willebrand disease registered within CNHP registry Annual Report 2022

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on behalf of

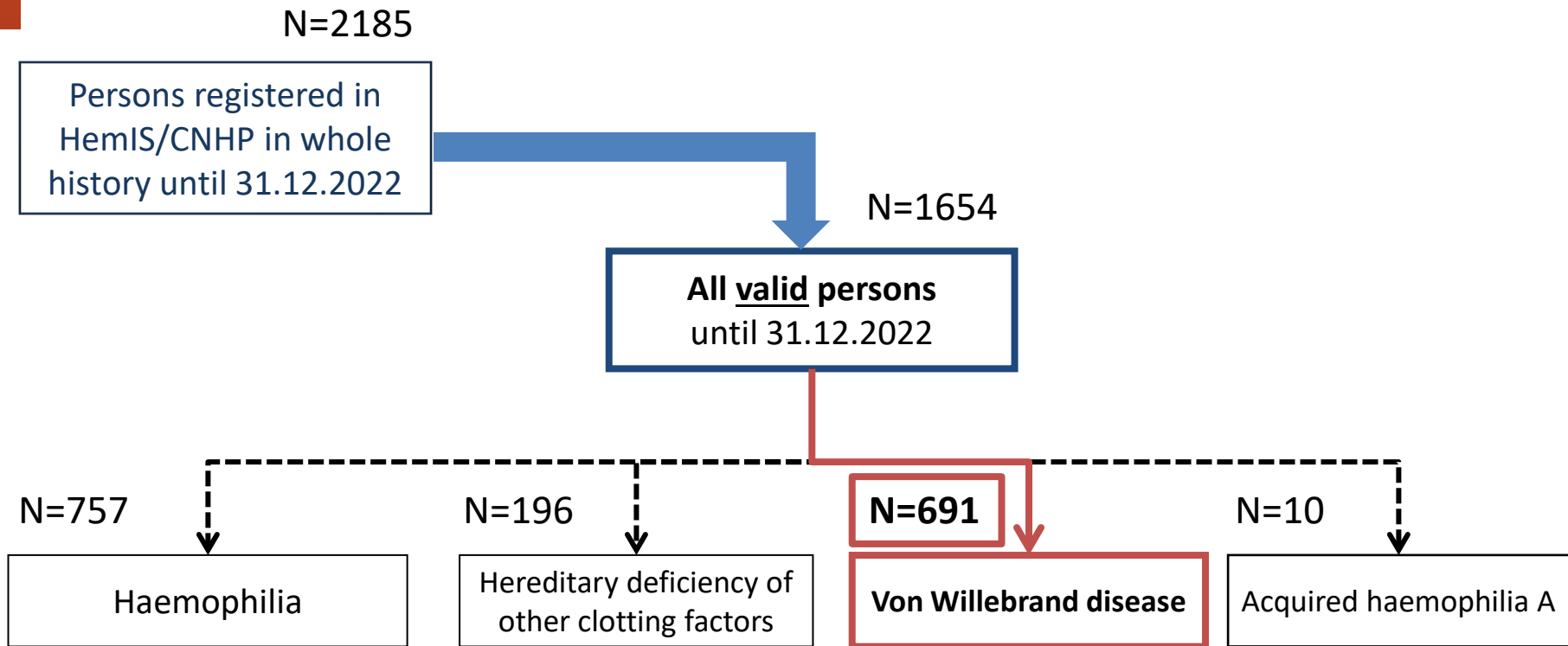
Centres contributing to database  
of the CNHP (Czech National Haemophilia Programme)

*Export date: March 28, 2023*

# Persons with Von Willebrand disease



# Sample size



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

# Number of patients in participating centres

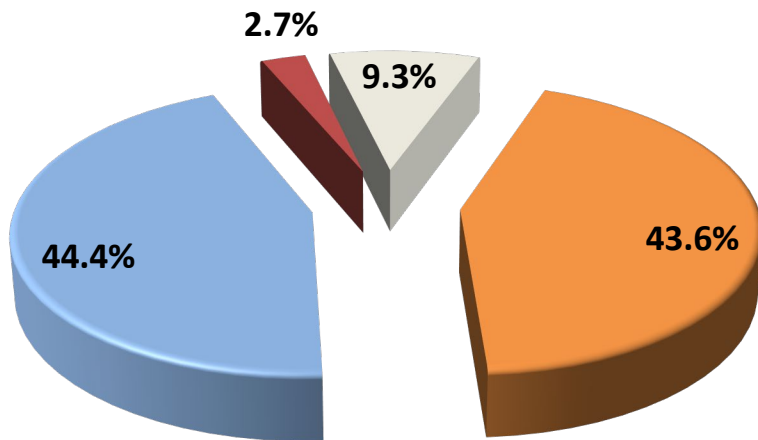
N=691





Paediatric centres	Valid patients	
	N	%
<b>Prague</b> – Dpt. of Pediatric Haematology and Oncology, CUH Motol	42	6.1
<b>Ostrava</b> – Dpt. of Pediatric Medicine, UH Ostrava	30	4.3
<b>Brno</b> – Dpt. of Pediatric Haematology, CUH Brno	27	3.9
<b>Pilsen</b> – Pediatric Dpt., UH Pilsen	21	3.0
<b>Hradec Králové</b> – Dpt. of Pediatric Medicine, UH HK	17	2.5
<b>Olomouc</b> – Dpt. of Pediatric Medicine, UH Olomouc	10	1.4
<b>Ústí n.L.</b> – Pediatric Dpt. – Haematology, Masaryk Hospital	9	1.3

Adult centres	Valid patients	
	N	%
<b>Brno</b> – Dpt. Of Clin Hematol, UH Brno	297	43.0
<b>Pilsen</b> – Dpt. of Biochemistry and Hematology, UH Pilsen – UKBH	81	11.7
<b>Ostrava</b> – Blood centre, UH Ostrava	72	10.4
<b>Liberec</b> – Dpt. Of Clin Hematol, Hospital Liberec	32	4.6
<b>Ústí n.L.</b> – Dpt. Of Clin Hematol, Masaryk Hospital	26	3.8
<b>Olomouc</b> – Haemato-Oncology Dpt., UH Olomouc	26	3.8
<b>České Budějovice</b> – Dpt. Of Clin Hematol, Hospital CB	1	0.1

# Type of Von Willebrand's disease

N=691



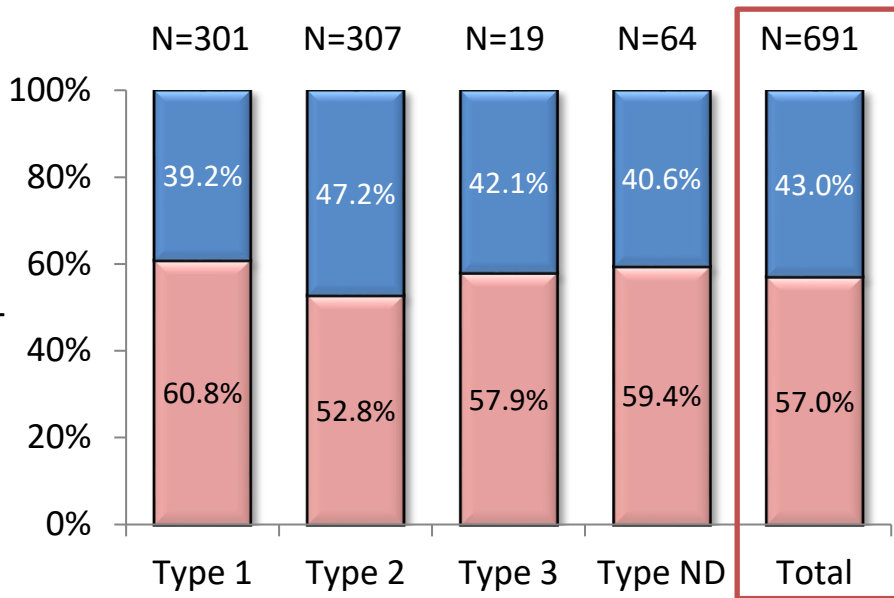
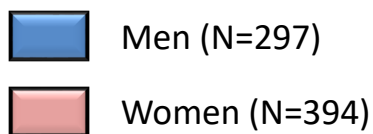
		Patients	
Type of disease		N	%
	Type 1*	301	43.6
	Type 2	307	44.4
	Type 2A	128	18.5
	Type 2B	19	2.7
	Type 2M	51	7.4
	Type 2N	30	4.3
	Type 2 (not specified)	79	11.4
	Type 3	19	2.7
	Type not determined / unfilled	64	9.3
Total		691	100.0

\* of them 7 patients with type 1/2N

# Sex and current age of patients

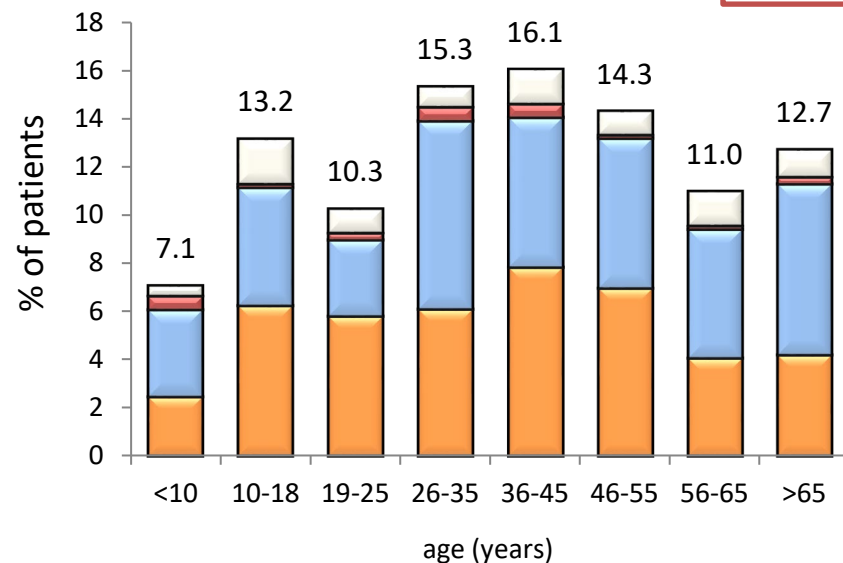
**N=691**

## Sex



## Current age\*

	Type 1	Type 2	Type 3	Type ND	Total
<b>N</b>	301	307	19	64	<b>691</b>
<b>Mean</b>	37.3	40.7	31.4	39.4	<b>38.8</b>
<b>Median</b>	37	39	29	39	<b>38</b>
<b>min - max</b>	2-94	1-86	0-71	1-84	<b>0-94</b>

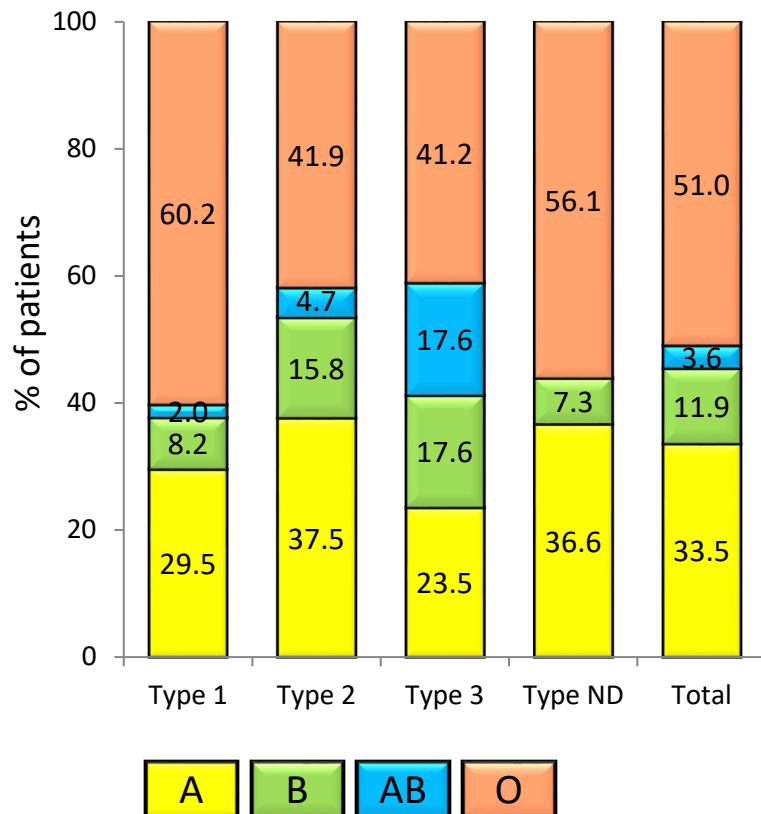


Type ND = not determined or unfilled

\* age reached in year 2022

# Blood group

N=691

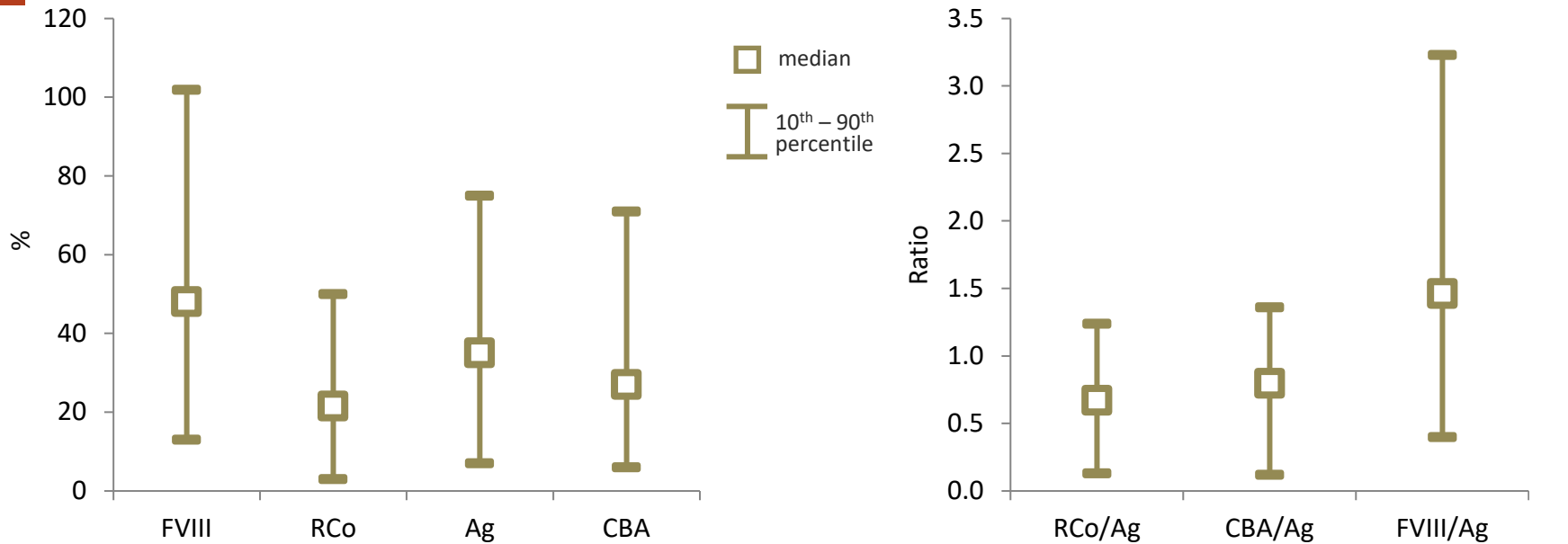


%<sup>1</sup> - % of total N  
 %<sup>2</sup> - % of valid N

Blood group	A	B	AB	O	ND	Valid N	Total N
<b>Type 1</b>	<i>N</i>	72	20	5	147	57	244
	% <sup>1</sup>	23.9	6.6	1.7	48.8	18.9	100.0
	% <sup>2</sup>	29.5	8.2	2.0	60.2		100.0
<b>Type 2</b>	<i>N</i>	95	40	12	106	54	253
	% <sup>1</sup>	30.9	13.0	3.9	34.5	17.6	100.0
	% <sup>2</sup>	37.5	15.8	4.7	41.9		100.0
<b>Type 3</b>	<i>N</i>	4	3	3	7	2	17
	% <sup>1</sup>	21.1	15.8	15.8	36.8	10.5	100.0
	% <sup>2</sup>	23.5	17.6	17.6	41.2		100.0
<b>Type ND</b>	<i>N</i>	15	3	0	23	23	41
	% <sup>1</sup>	23.4	4.7	0.0	35.9	35.9	100.0
	% <sup>2</sup>	36.6	7.3	0.0	56.1		100.0
<b>Total</b>	<i>N</i>	186	66	20	283	136	555
	% <sup>1</sup>	26.9	9.6	2.9	41.0	19.7	100.0
	% <sup>2</sup>	33.5	11.9	3.6	51.0		100.0

# Factor levels

N=691



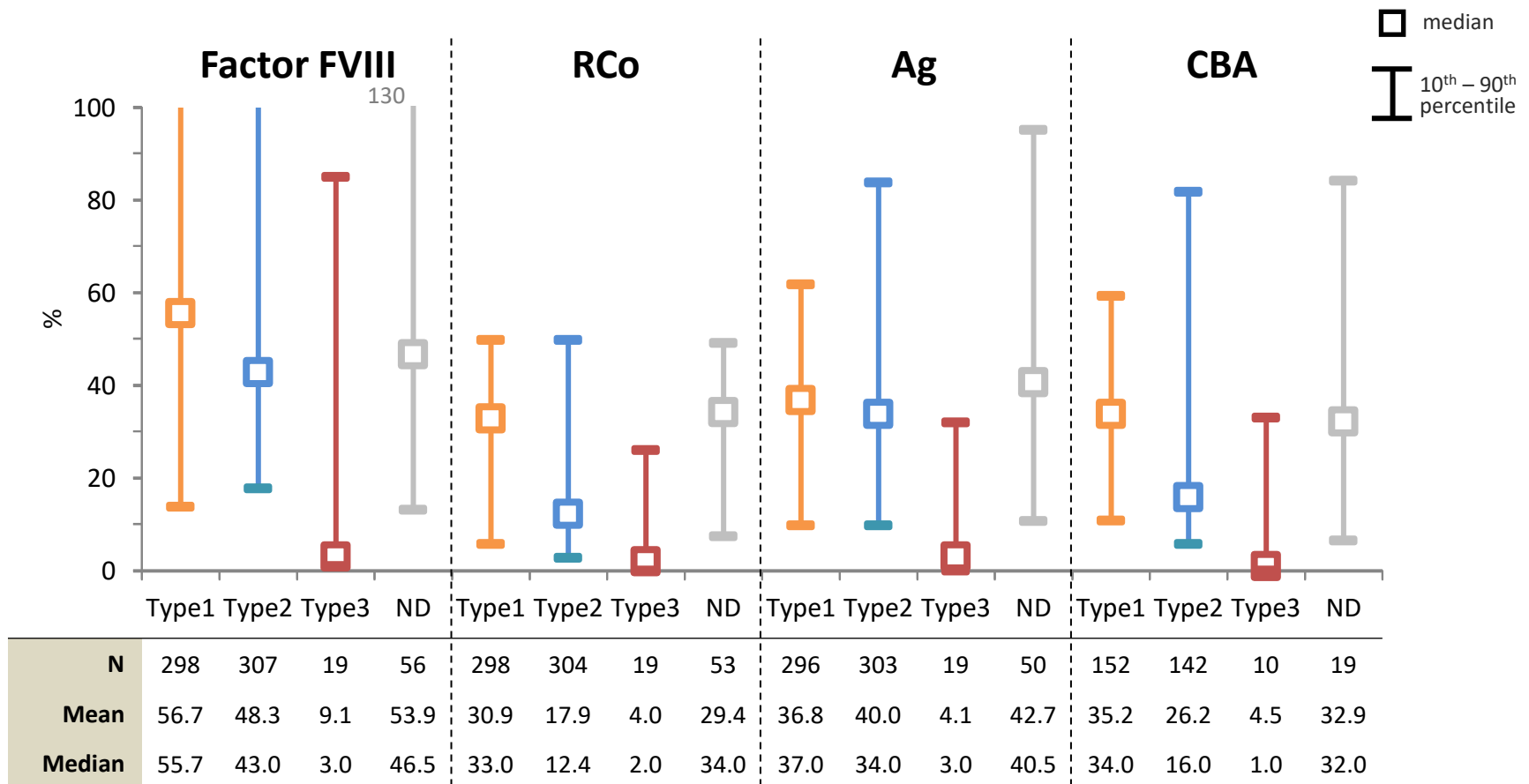
Factor FVIII*	RCo	Ag	CBA		RCo/Ag	CBA/Ag	FVIII/Ag
680	674	668	323	<b>N</b>	660	322	663
51.4	24.2	37.8	30.2	<b>Mean</b>	0.8	0.8	1.8
48 (1–232)	21.5 (0–94)	35 (0–261)	27 (0–127)	<b>Median (min – max)</b>	0.67 (0–38)	0.795 (0–3)	1.46 (0.08–106)

\* Factor FVIII was assessed by coagulation method in 627 patients, by chromogenic method in 20 patients and method is missing in 33 patients.



# Factor levels according to type of VWD, part 1

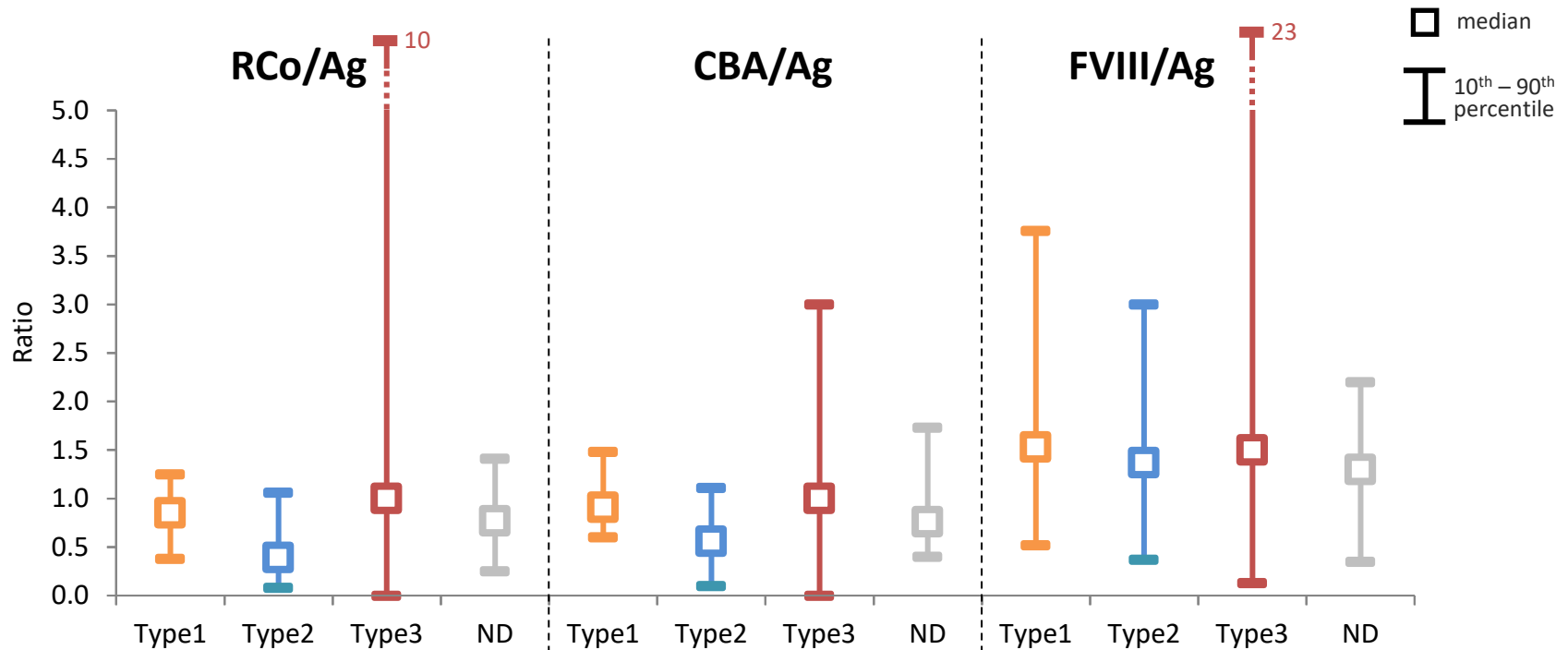
N=691



Type ND = not determined or unfilled

# Factor levels according to type of VWD, part 2

N=691



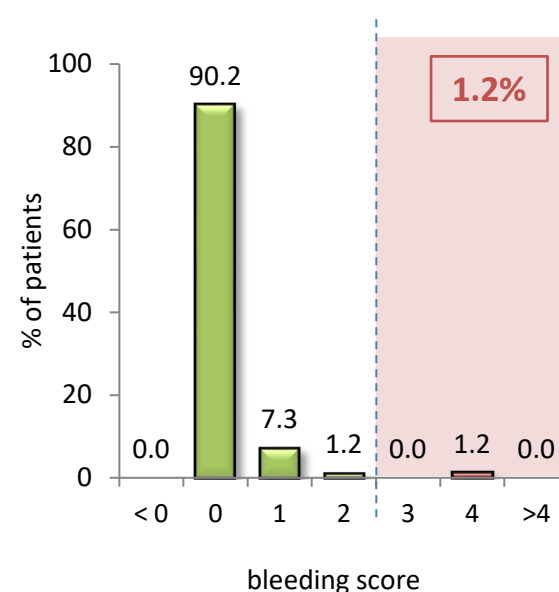
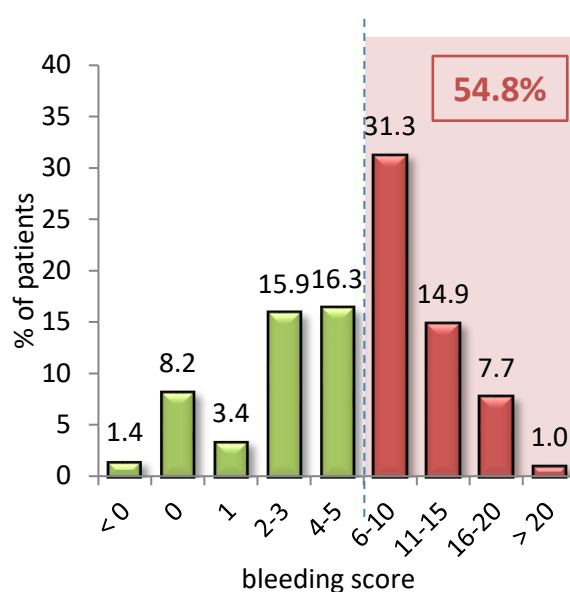
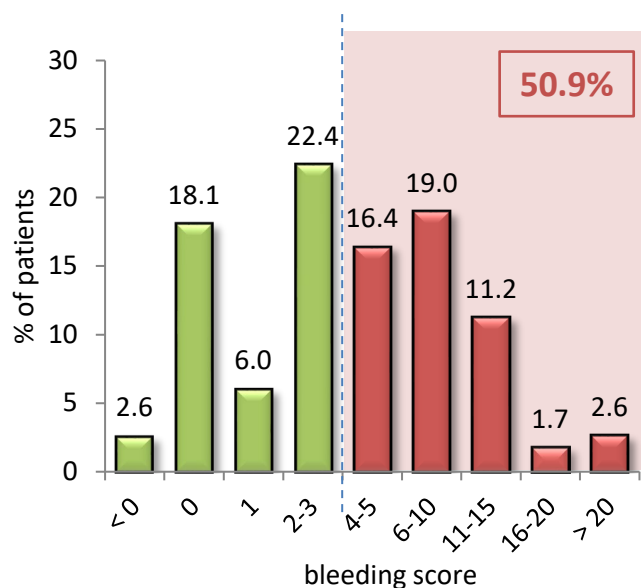
	Type1	Type2	Type3	ND	Type1	Type2	Type3	ND	Type1	Type2	Type3	ND
<b>N</b>	295	299	17	49	151	142	10	19	295	301	17	50
<b>Mean</b>	1.02	0.46	1.41	0.79	0.96	0.57	0.90	0.84	2.17	1.52	3.33	1.38
<b>Median</b>	0.85	0.39	1.00	0.77	0.91	0.56	1.00	0.76	1.53	1.37	1.50	1.30

Type ND = not determined or unfilled

# Bleeding score<sup>1</sup> according to sex and age

N=406<sup>2</sup>

	Adult men	Adult women	Children
<b>N</b>	116	208	82
<b>Mean</b>	5.1	7.0	0.1
<b>Median (min - max)</b>	4 (-2 - 25)	6 (-2 - 24)	0 (0 - 4)



<sup>1</sup> Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

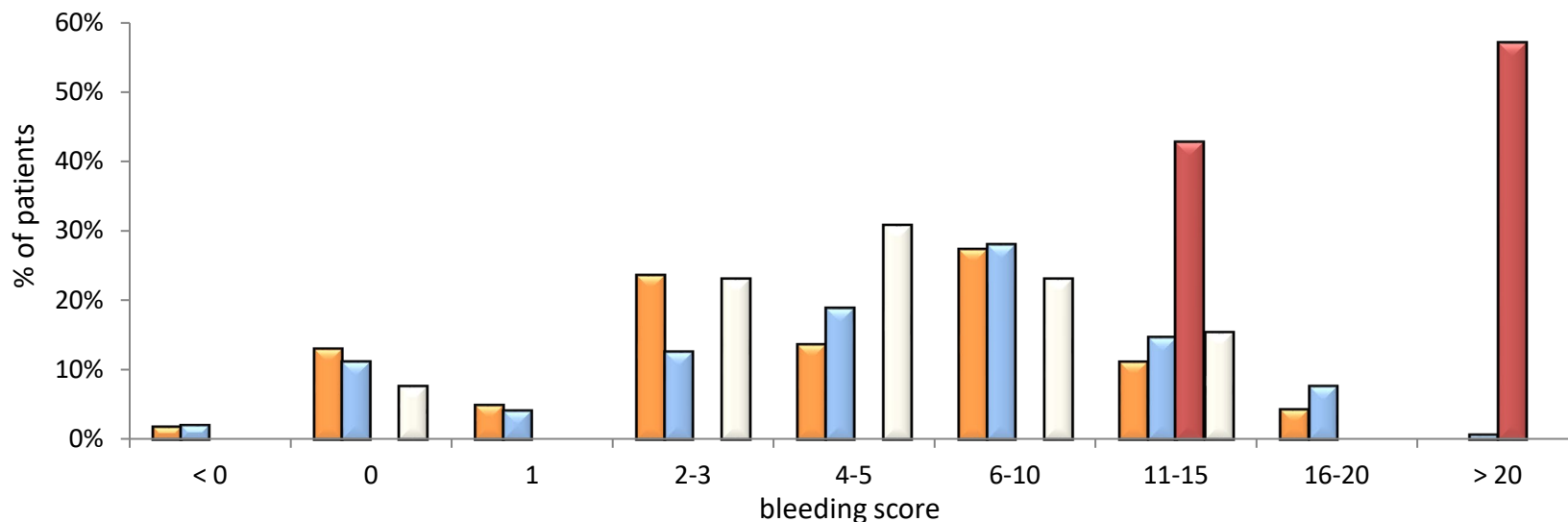
<sup>2</sup> Missing information on bleeding score in 285 patients.

# Bleeding score<sup>1</sup> in adults according to type of disease

N=324<sup>2</sup>

Bleeding score

	Type 1	Type 2	Type 3	Type ND	Total
<b>N total</b>	241	248	14	48	<b>551</b>
<b>N valid</b>	161	143	7	13	<b>324</b>
<b>Mean</b>	<b>5.4</b>	<b>6.7</b>	<b>18.9</b>	<b>5.8</b>	<b>6.3</b>
<b>Median</b>	<b>5.0</b>	<b>6.0</b>	<b>21.0</b>	<b>5.0</b>	<b>5.0</b>
<b>min - max</b>	-2-20	-1-22	13-25	0-14	<b>-2-25</b>



<sup>1</sup> Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

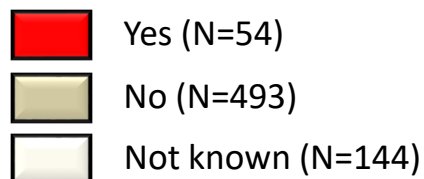
<sup>2</sup> Adult patients with non-missing information on bleeding score.

Type ND = not determined or unfilled

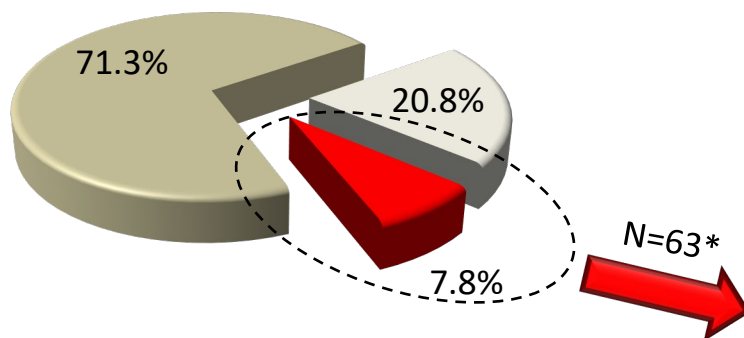
# Concomitant diseases

N=691

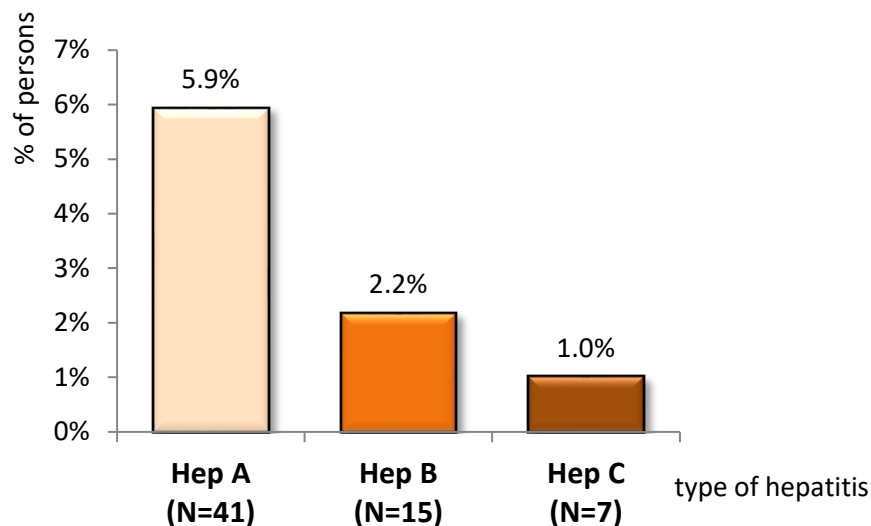
## Experienced hepatitis



None of the patients is HIV positive.



\*Total of 63 cases of hepatitis in 54 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=691
<b>ABR</b>						
N valid	301	307	19	64	691	
Mean	0.04	0.09	1.21	0.00	0.09	
Median	0	0	1	0	0	
min-max	0-3	0-8	0-6	0-0	0-8	
SUM	11	27	23	0	61	
<hr/>						
<b>Location of bleeds</b>						
$N_p/N_B^2$						
Joints	1/2	2/2	2/3	0/-	5/7	
Epistaxes	1/2	5/12	5/13	0/-	11/27	
Urogenital tract	1/1	3/3	1/1	0/-	5/5	
GIT	-	-	-	-	-	
Subcutaneous	1/1	3/3	1/1	0/-	5/5	
Muscles	1/1	2/2	1/1	0/-	4/4	
CNS	1/1	0/-	1/1	0/-	2/2	
Metrorrhagia	1/2	0/-	0/-	0/-	1/2	
Other	1/1	2/2	1/1	0/-	4/4	
<b>SUM</b>	<b>8/11</b>	<b>17/24</b>	<b>12/21</b>	<b>0/-</b>	<b>37/56</b>	

<sup>2</sup>  $N_p$  = 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 2022

N=691

	Number of patients treated with substitution of VWF total (type1/type2/type3/typeND)	Total annual consumption (IU)	Average annual consumption per treated patient (IU)
<b>Haemate P</b>	<b>68 (18/39/9/2)</b>	<b>1 687 291</b>	<b>24 813.1</b>
<i>of them on prophylaxis*</i>	<i>4 (0/0/4/0)</i>	<i>1 042 000</i>	<i>260 500.0</i>
<b>Wilate</b>	<b>29 (16/10/3/0)</b>	<b>571 350</b>	<b>19 701.7</b>
<i>of them on prophylaxis*</i>	<i>2 (0/0/2/0)</i>	<i>203 500</i>	<i>101 750.0</i>
<b>Fanhdi</b>	<b>6 (3/3/0/0)</b>	<b>71 000</b>	<b>11 833.3</b>
<i>of them on prophylaxis*</i>	<i>1 (0/1/0/0)</i>	<i>51 000</i>	<i>51 000.0</i>
<b>Total</b>	<b>103 (37/52/12/2)</b>	<b>2 329 641</b>	<b>22 617.9</b>
<i>of them on prophylaxis*</i>	<i>7 (0/1/6/0)</i>	<i>1 296 500</i>	<i>185 214.3</i>
Total - type 1	37 (37/0/0/0)	177 000	<u>4 783.8</u>
Total - type 2	52 (0/52/0/0)	754 291	<u>14 505.6</u>
Total - type 3	12 (0/0/12/0)	1 364 350	<u>113 695.8</u>
Total - type ND	2 (0/0/0/2)	34 000	<u>17 000.0</u>

\* permanent prophylaxis

Type ND = not determined or unfilled

# Consumption of other drugs in year 2022

N=691

	Number of patients treated with other drugs total (type1/type2/type3/typeND)	Total annual consumption	Average annual consumption per <u>treated</u> patient
<b>DDAVP (µg)</b>	<b>2 (0/2/0/0)</b>	<b>35</b>	<b>17.6</b>
<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=7

No	Centre	Age group	Type of VWD	Type of prophylaxis in 2022	Treatment in 2022	No of applications per week	Total consumption (IU)	ABR in 2022
1	Praha	Child	3	<b>Permanent</b>	Wilate	3	151 000	5
2	Ostrava	Child	3	<b>Permanent</b>	Wilate	2	52 500	1
3	Brno	Adult	3	<b>Permanent</b>	Haemate P	2.3	384 000	6
4	Brno	Adult	3	<b>Permanent</b>	Haemate P	2	250 000	1
5	Brno	Adult	3	<b>Permanent</b>	Haemate P	2	220 000	3
6	Brno	Adult	3	<b>Permanent</b>	Haemate P	2	188 000	0
7	Plzeň	Adult	2	<b>Permanent</b>	Fanhdi	NA	51 000	0