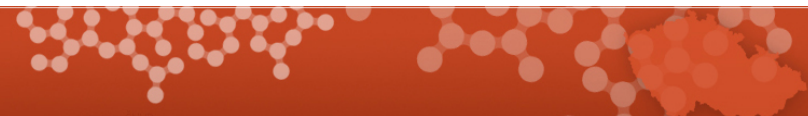


# **The status of care for persons with von Willebrand disease registered within CNHP registry Annual Report 2020**

Jan Blatný, Petra Ovesná, Karolína Hurdálková  
on behalf of

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

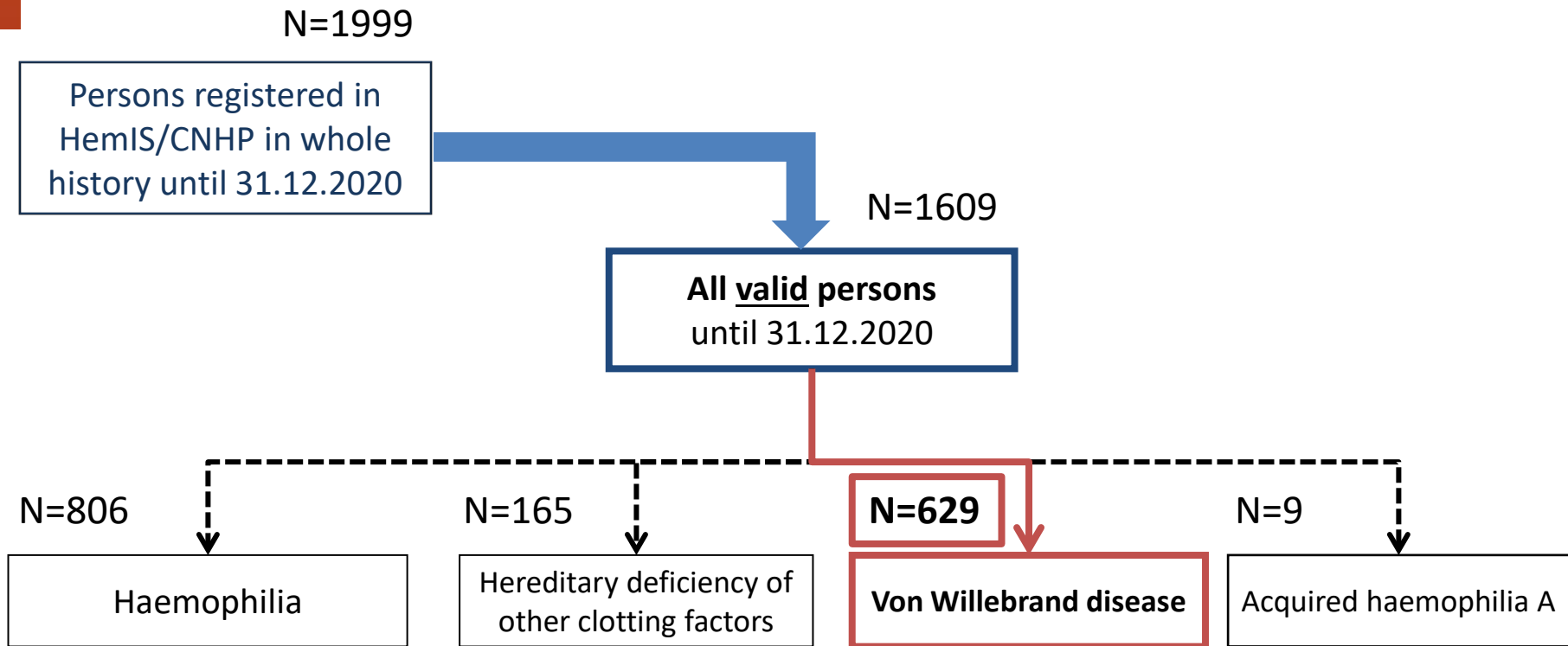
*Export date: March 30, 2021*



# Persons with Von Willebrand disease



# Sample size



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

# Number of patients in participating centres

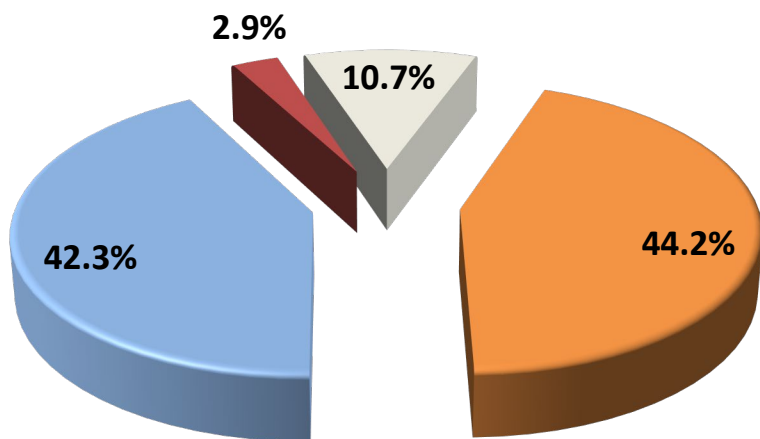
N=629





Paediatric centres	Valid patients	
	N	%
<b>Prague</b> – Dpt. of Pediatric Haematology and Oncology, CUH Motol	28	4.5
<b>Pilsen</b> – Pediatric Dpt., UH Pilsen	24	3.8
<b>Ostrava</b> – Dpt. of Pediatric Medicine, UH Ostrava	24	3.8
<b>Hradec Králové</b> – Dpt. of Pediatric Medicine, UH HK	20	3.2
<b>Brno</b> – Dpt. of Pediatric Haematology, CUH Brno	16	2.5
<b>Ústí n.L.</b> – Pediatric Dpt. – Haematology, Masaryk Hospital	12	1.9
<b>Olomouc</b> – Dpt. of Pediatric Medicine, UH Olomouc	1	0.2

Adult centres	Valid patients	
	N	%
<b>Brno</b> – Dpt. Of Clin Hematol, UH Brno	283	45.0
<b>Pilsen</b> – Dpt. of Biochemistry and Hematology, UH Pilsen – UKBH	77	12.2
<b>Ostrava</b> – Blood centre, UH Ostrava	71	11.3
<b>Liberec</b> – Dpt. Of Clin Hematol, Hospital Liberec	28	4.5
<b>Olomouc</b> – Haemato-Oncology Dpt., UH Olomouc	23	3.7
<b>Ústí n.L.</b> – Dpt. Of Clin Hematol, Masaryk Hospital	18	2.9
<b>Hradec Králové</b> – IV. Internal and Hematology Dpt., UH HK	2	0.3
<b>České Budějovice</b> – Dpt. Of Clin Hematol, Hospital CB	1	0.2

# Type of Von Willebrand's disease

N=629



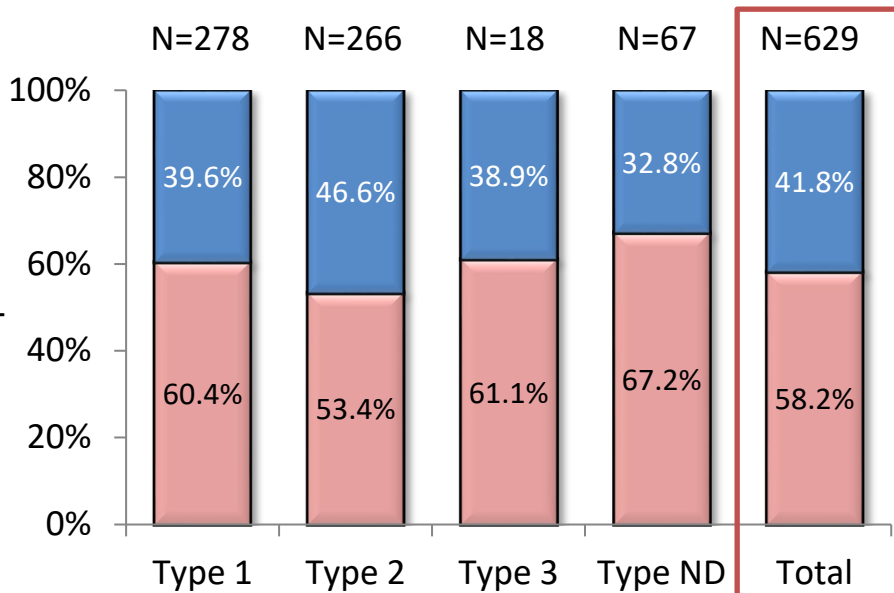
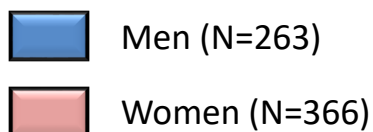
		Patients	
Type of disease		N	%
	Type 1*	278	44.2
	Type 2	266	42.3
	Type 2A	113	18.0
	Type 2B	20	3.2
	Type 2M	44	7.0
	Type 2N	24	3.8
	Type 2 (not specified)	65	10.3
	Type 3	18	2.9
	Type not determined / unfilled	67	10.7
Total		629	100.0

\* of them 6 patients with type 1/2N

# Sex and current age of patients

**N=629**

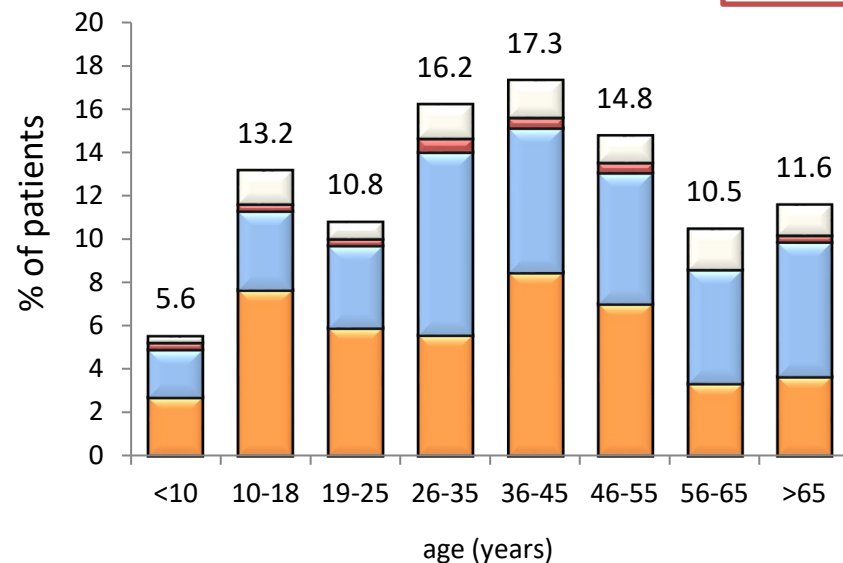
## Sex



Type ND = not determined or unfilled

## Current age\*

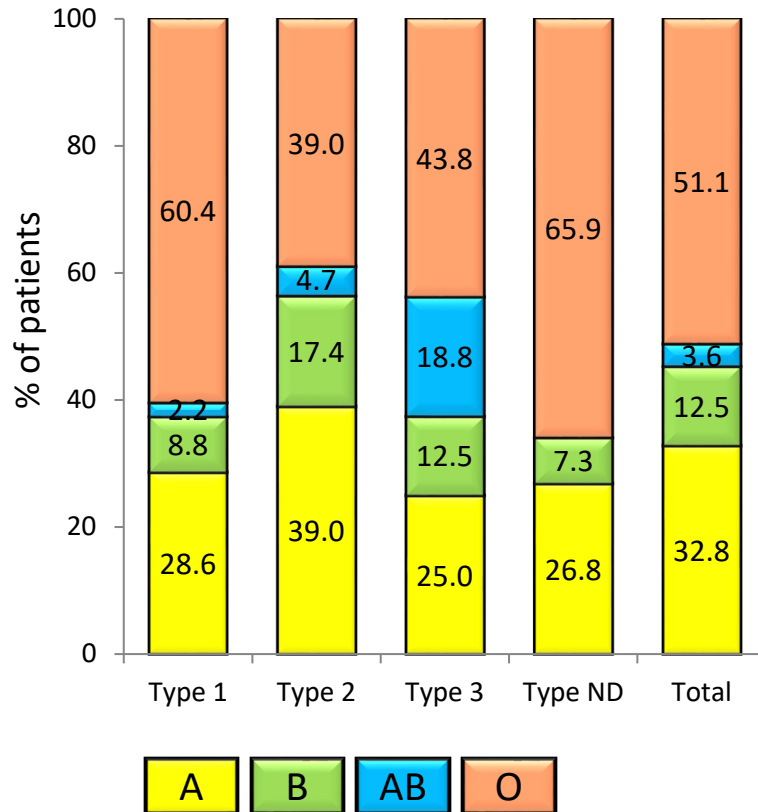
	Type 1	Type 2	Type 3	Type ND	Total
<b>N</b>	278	266	18	67	<b>629</b>
<b>Mean</b>	36.2	41.6	33.6	42.7	<b>39.1</b>
<b>Median</b>	36	41	33.5	42	<b>38</b>
<b>min - max</b>	3 - 92	3 - 87	1 - 69	7 - 83	<b>1 - 92</b>



\* age reached in year 2020

# Blood group

N=629

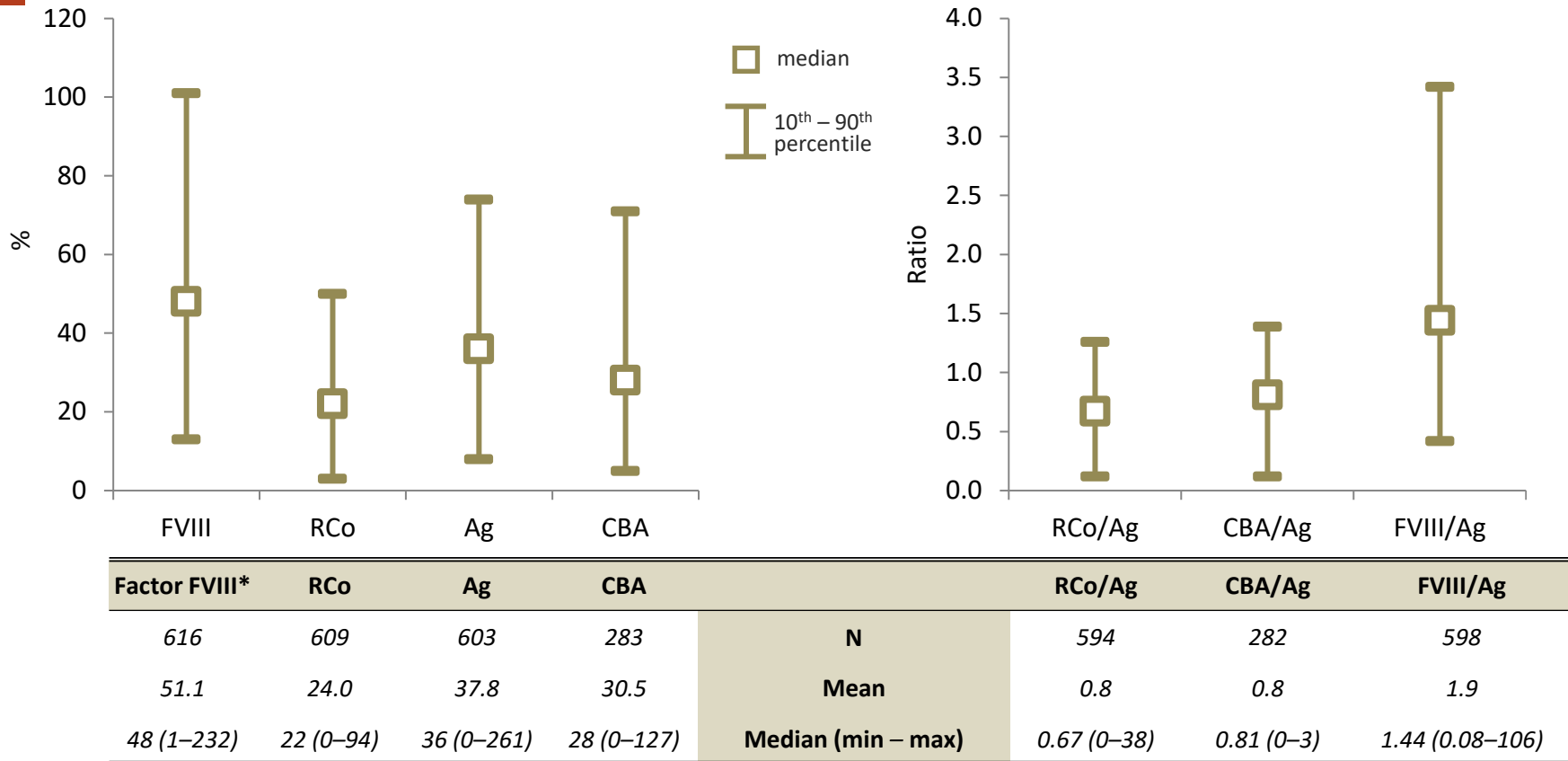


%<sup>1</sup> - % of total N  
 %<sup>2</sup> - % of valid N

Blood group	A	B	AB	O	ND	Valid N	Total N
<b>Type 1</b>	<i>N</i>	65	20	5	137	51	227
	% <sup>1</sup>	23.4	7.2	1.8	49.3	18.3	100.0
	% <sup>2</sup>	28.6	8.8	2.2	60.4		100.0
<b>Type 2</b>	<i>N</i>	83	37	10	83	53	213
	% <sup>1</sup>	31.2	13.9	3.8	31.2	19.9	100.0
	% <sup>2</sup>	39.0	17.4	4.7	39.0		100.0
<b>Type 3</b>	<i>N</i>	4	2	3	7	2	16
	% <sup>1</sup>	22.2	11.1	16.7	38.9	11.1	100.0
	% <sup>2</sup>	25.0	12.5	18.8	43.8		100.0
<b>Type ND</b>	<i>N</i>	11	3	0	27	26	41
	% <sup>1</sup>	16.4	4.5	0.0	40.3	38.8	100.0
	% <sup>2</sup>	26.8	7.3	0.0	65.9		100.0
<b>Total</b>	<i>N</i>	163	62	18	254	132	497
	% <sup>1</sup>	25.9	9.9	2.9	40.4	21.0	100.0
	% <sup>2</sup>	32.8	12.5	3.6	51.1		100.0

# Factor levels

N=629

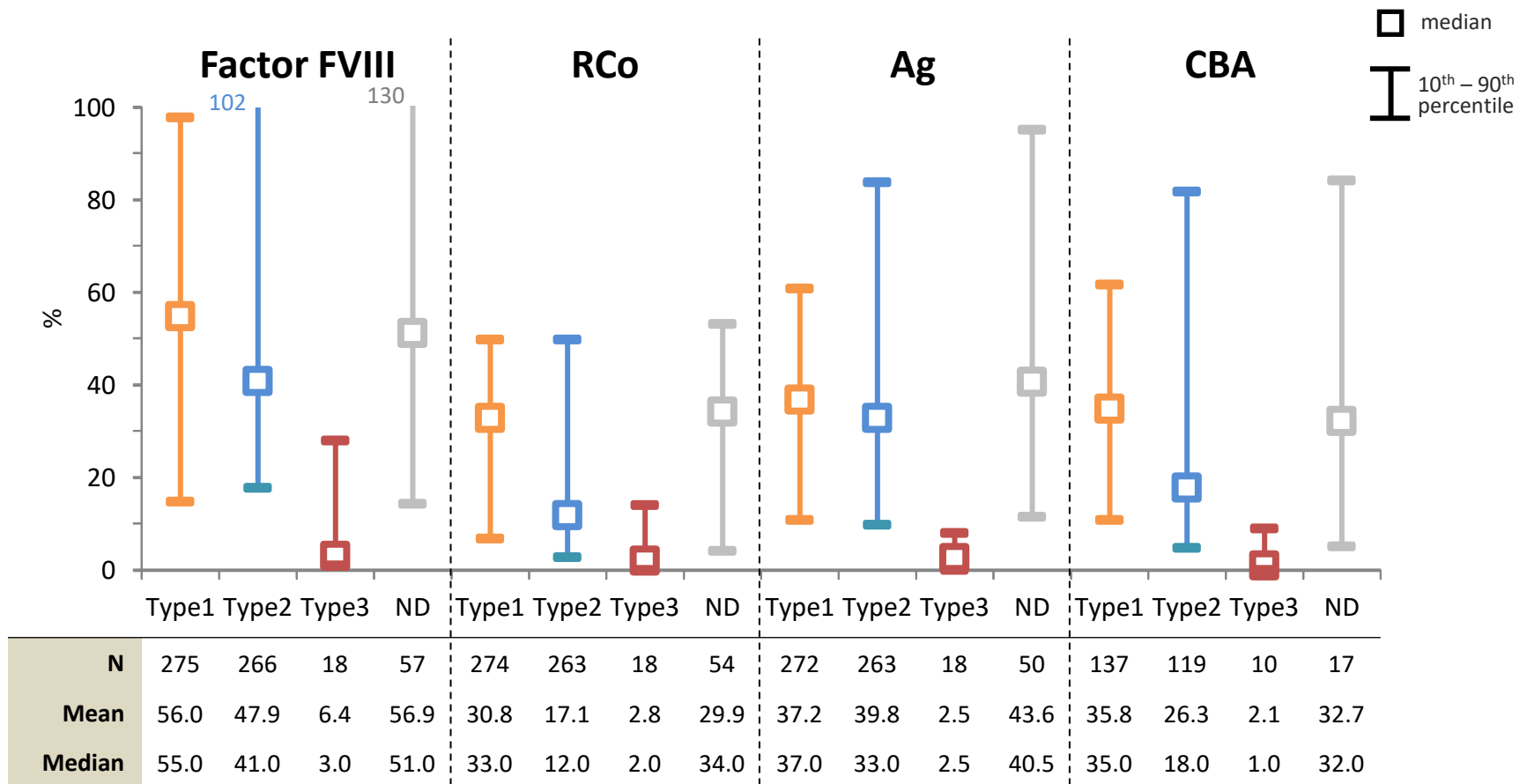


\* Factor FVIII was assessed by coagulation method in 571 patients, by chromogenic method in 18 patients and method is missing in 40 patients.



# Factor levels according to type of VWD, part 1

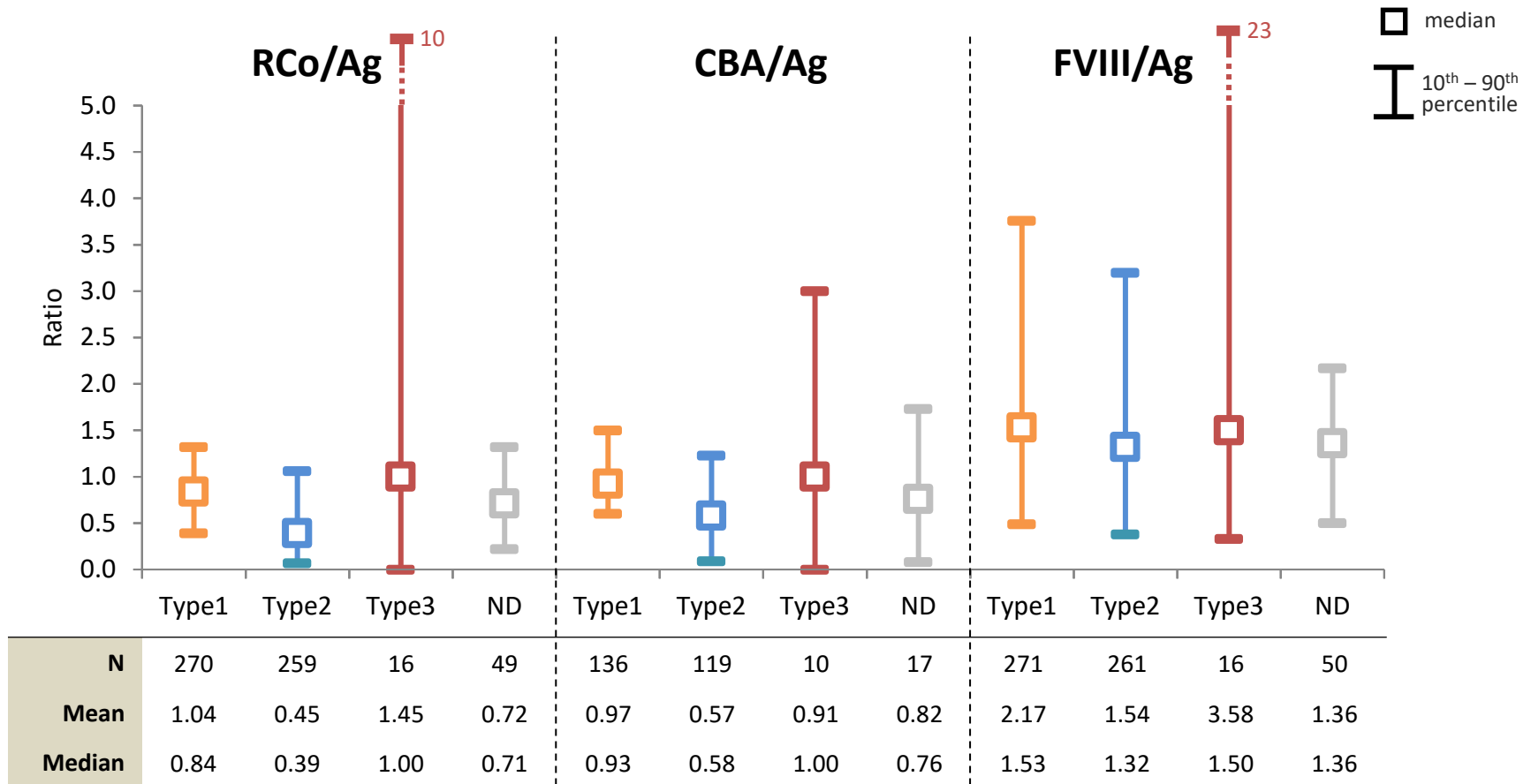
N=629



Type ND = not determined or unfilled

# Factor levels according to type of VWD, part 2

N=629

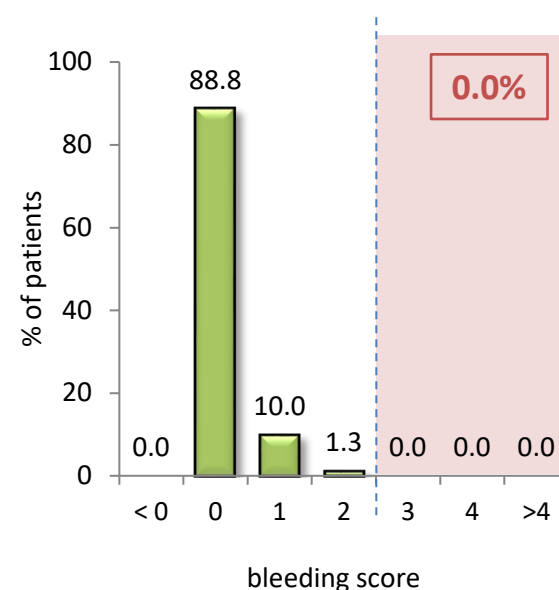
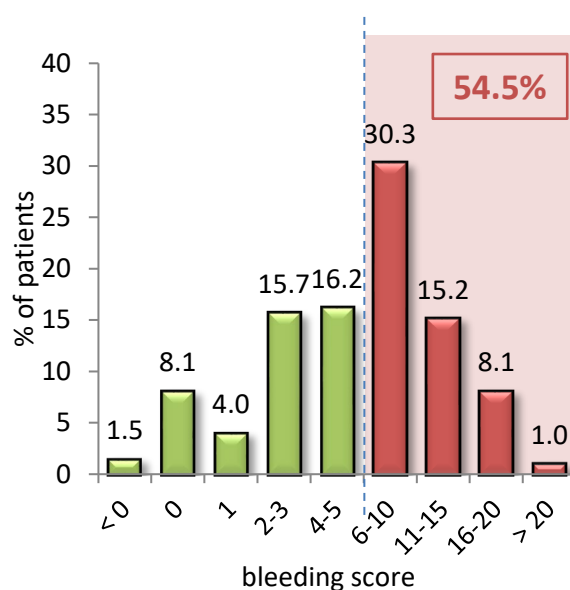
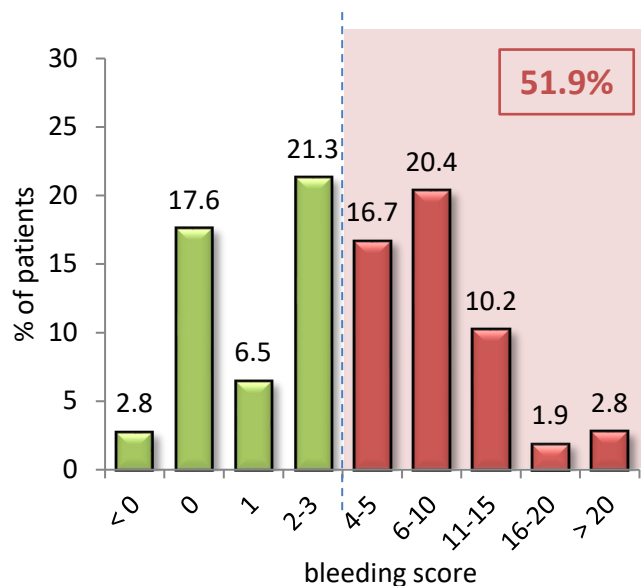


Type ND = not determined or unfilled

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

N=386<sup>2</sup>

	Adult men	Adult women	Children
<b>N</b>	108	198	80
<b>Mean</b>	5.1	7.0	0.1
<b>Median (min - max)</b>	4 (-2 - 25)	6 (-2 - 24)	0 (0 - 2)



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

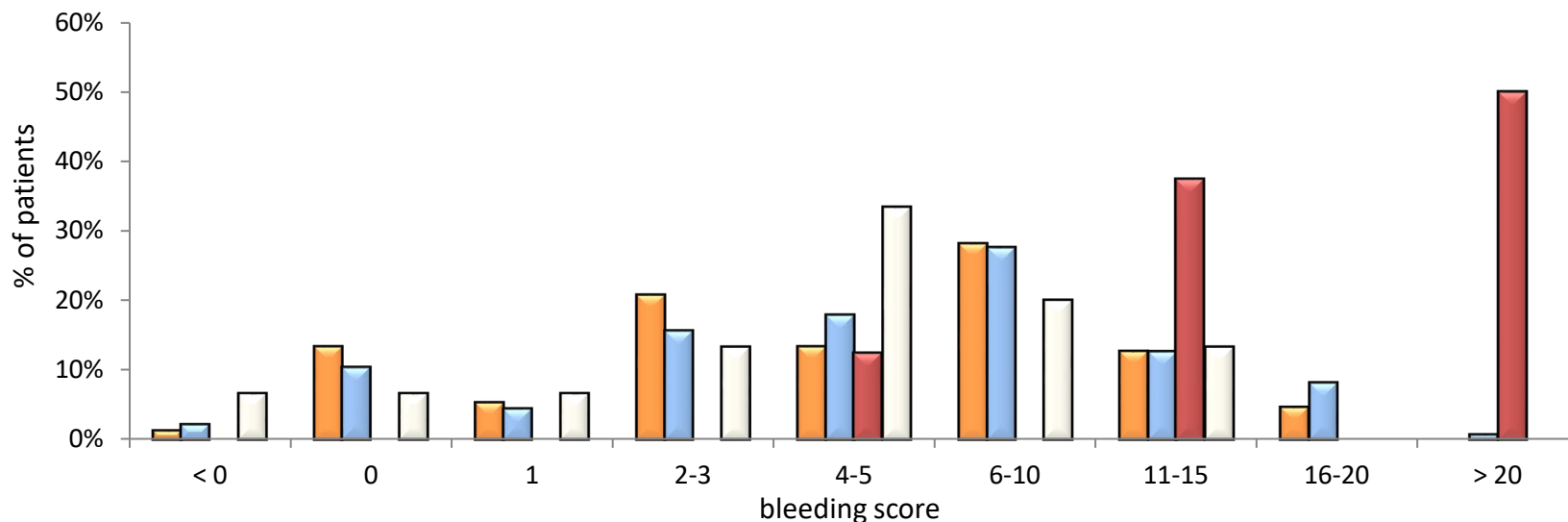
<sup>2</sup> Missing information on bleeding score in 243 patients.

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

N=306<sup>2</sup>

Bleeding score

	Type 1	Type 2	Type 3	Type ND	Total
<b>N total</b>	213	229	14	55	<b>511</b>
<b>N valid</b>	149	134	8	15	<b>306</b>
<b>Mean</b>	<b>5.6</b>	<b>6.6</b>	<b>17.1</b>	<b>5.3</b>	<b>6.3</b>
<b>Median</b>	<b>5.0</b>	<b>5.0</b>	<b>17.5</b>	<b>5.0</b>	<b>5.0</b>
<b>min - max</b>	-2-20	-1-22	5-25	-2-14	<b>-2-25</b>



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

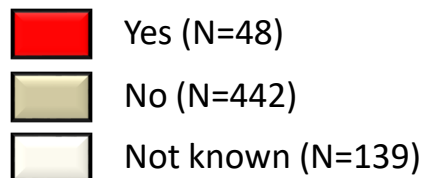
<sup>2</sup> Adult patients with non-missing information on bleeding score.

Type ND = not determined or unfilled

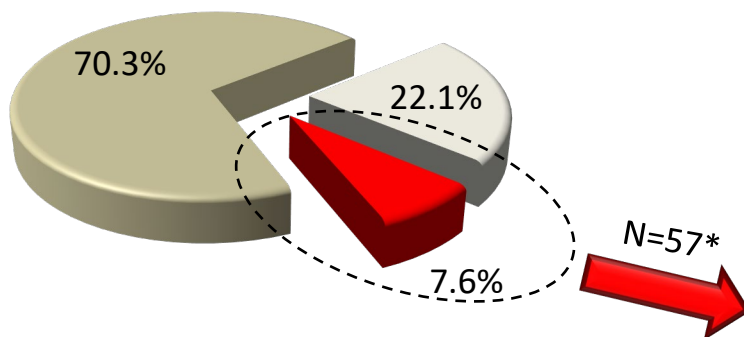
# Concomitant diseases

N=629

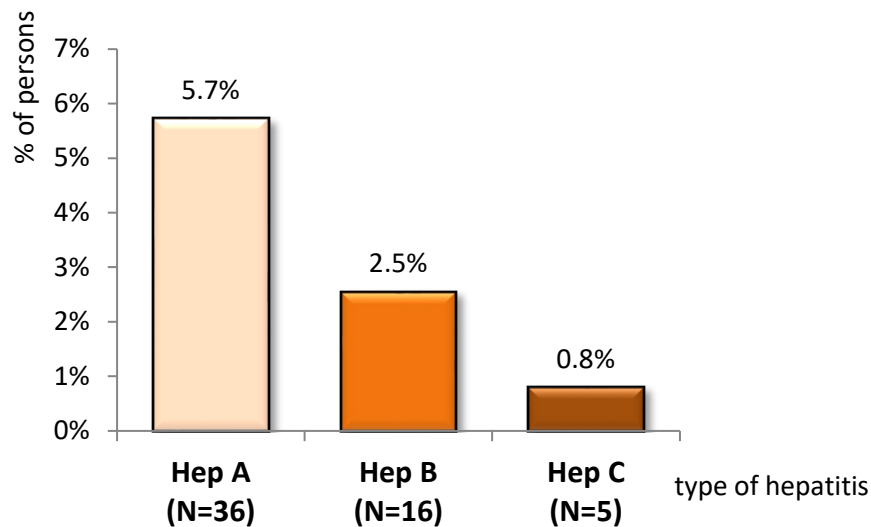
## Experienced hepatitis



None of the patients is HIV positive.



\*Total of 57 cases of hepatitis in 48 persons. One person may have more types of hepatitis recorded.



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

**N=629**

	Type 1	Type 2	Type 3	Type ND	Total
<b>ABR</b>					
<b>N valid</b>	278	266	18	67	629
<b>Mean</b>	0.01	0.07	1.22	0.07	0.07
<b>Median</b>	0	0	0	0	0
<b>min-max</b>	0-1	0-6	0-7	0-4	0-7
<b>SUM</b>	2	18	22	5	47
<hr/>					
<b>Location of bleeds</b>					
<b><math>N_p/N_B^2</math></b>					
<b>Joints</b>	0/-	1/1	2/8	0/-	3/9
<b>Epistaxes</b>	1/1	3/5	2/5	1/4	7/15
<b>Urogenital tract</b>	0/-	/-	/-	0/-	/-
<b>GIT</b>	0/-	2/2	0/-	0/-	2/2
<b>Subcutaneous</b>	0/-	3/3	2/2	0/-	5/5
<b>Muscles</b>	1/1	1/1	2/2	0/-	4/4
<b>CNS</b>	0/-	0/-	0/-	0/-	0/-
<b>Metrorrhagia</b>	0/-	0/-	1/2	0/-	1/2
<b>Other</b>	0/-	3/3	3/3	1/1	7/7
<b>SUM</b>	<b>2/2</b>	<b>13/15</b>	<b>12/22</b>	<b>2/5</b>	<b>29/44</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 2020

N=629

	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>Fanhdi</b>	<b>11 (3/7/0/1)</b>	<b>136 000</b>	<b>12 363.6</b>
<i>of them on prophylaxis*</i>	<i>1 (0/1/0/0)</i>	<i>51 000</i>	<i>51 000.0</i>
<b>Haemate P</b>	<b>61 (9/40/9/3)</b>	<b>1 384 961</b>	<b>22 704.3</b>
<i>of them on prophylaxis*</i>	<i>2 (0/0/2/0)</i>	<i>487 000</i>	<i>243 500.0</i>
<b>Wilate</b>	<b>22 (10/6/5/1)</b>	<b>284 250</b>	<b>12 920.5</b>
<i>of them on prophylaxis*</i>	<i>5 (0/0/5/0)</i>	<i>177 500</i>	<i>35 500.0</i>
<b>Total</b>	<b>94 (22/53/14/5)</b>	<b>1 805 211</b>	<b>18 056.7</b>
<i>of them on prophylaxis*</i>	<i>8 (0/1/7/0)</i>	<i>715 500</i>	<i>89 437.5</i>
Total - type 1	22	176 750	<u>8 034.1</u>
Total - type 2	53	630 923	<u>11 712.0</u>
Total - type 3	14	919 511	<u>65 679.4</u>
Total - type ND	5	78 027	<u>13 004.5</u>

\* permanent prophylaxis

Type ND = not determined or unfilled

# Consumption of other drugs in year 2020

N=629

	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 (0/1/0/1)</b>	<b>1 527</b>	<b>763.5</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=6

No	Centre	Age group	Type of VWD	Type of prophylaxis in 2020	Treatment in 2020	No of applications per week	Total consumption (IU)	ABR in 2020
1	Plzeň	Adult	2	<b>Permanent</b>	Fanhdi	1	51 000	0
2	Praha	Child	3	<b>Permanent</b>	Wilate	2	114 000	6
3	Ostrava	Child	3	<b>Permanent</b>	Wilate	2	23 500	1
4	Brno	Adult	3	<b>Permanent</b>	Haemate P	2	305 000	7
5	Brno	Adult	3	<b>Permanent</b>	Haemate P	2	182 000	0
6	Olomouc	Adult	3	<b>Permanent</b>	Wilate	1	40 000	0