

The status of care for persons with von Willebrand disease registered within CNHP registry Annual Report 2017

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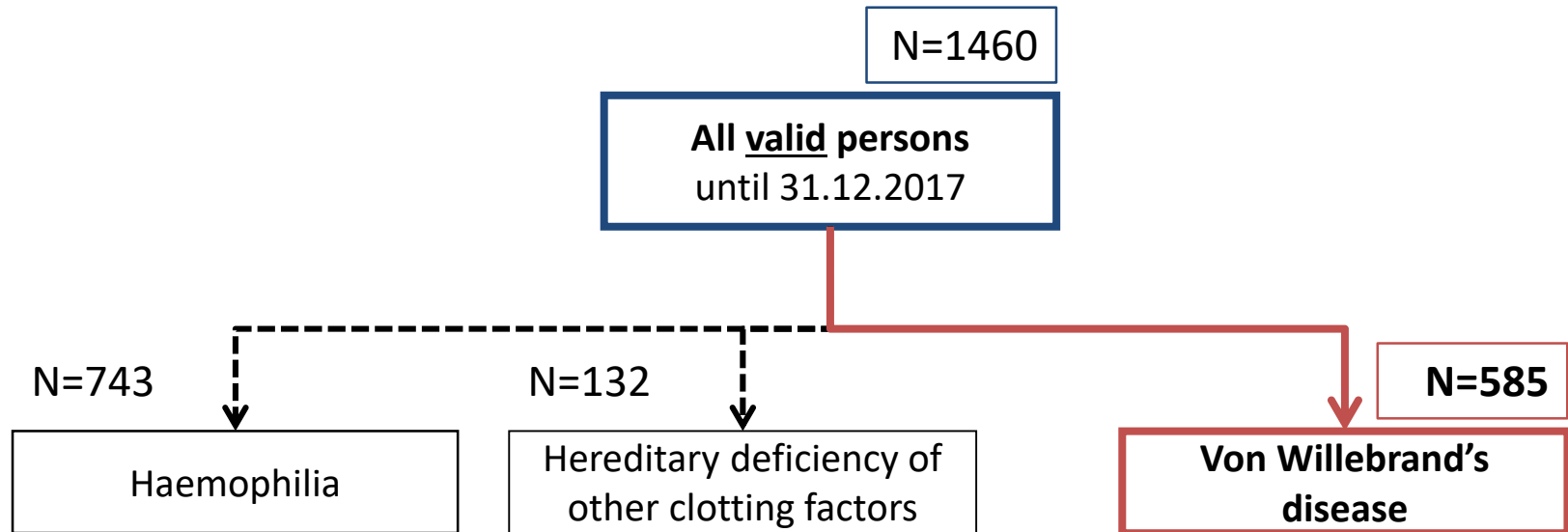
on behalf of

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

Export date: April 2018

Persons with Von Willebrand disease

Sample size



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

Number of patients in participating centres

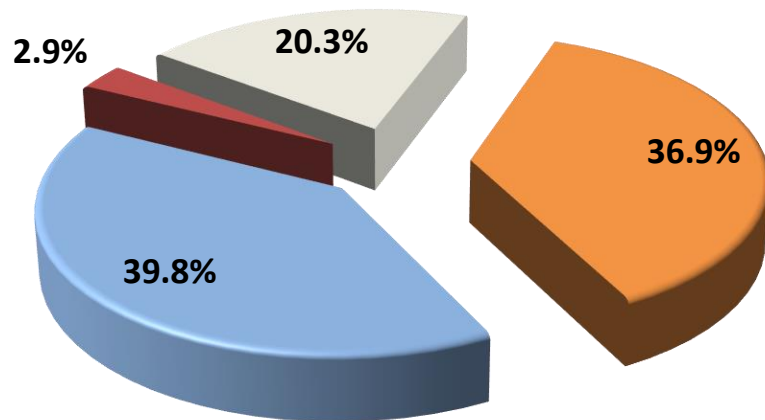
N=585

Paediatric centres	Valid patients	
	N	%
Brno – Dpt. of Pediatric Haematology, CUH Brno	27	4.6
Prague – Dpt. of Pediatric Haematology and Oncology, CUH Motol	26	4.4
Pilsen – Pediatric Dpt., UH Pilsen	22	3.8
Hradec Králové – Dpt. of Pediatric Medicine, UH HK	17	2.9
Ústí n.L. – Pediatric Dpt. – Haematology, Masaryk Hospital	15	2.6
Ostrava – Dpt. of Pediatric Medicine, UH Ostrava	14	2.4
Olomouc – Dpt. of Pediatric Medicine, UH Olomouc	1	0.2

Adult centres	Valid patients	
	N	%
Brno – Dpt. Of Clin Hematol, UH Brno	272	46.5
Pilsen – Dpt. of Biochemistry and Hematology, UH Pilsen – UKBH	74	12.6
Ostrava – Blood centre, UH Ostrava	69	11.8
Liberec – Dpt. Of Clin Hematol, Hospital Liberec	27	4.6
Olomouc – Haemato-Oncology Dpt., UH Olomouc	13	2.2
Ústí n.L. – Dpt. Of Clin Hematol, Masaryk Hospital	5	0.9
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=585



Type of disease	Patients	
	N	%
Type 1*	216	36.9
Type 2	233	39.8
Type 2A	103	17.6
Type 2B	17	2.9
Type 2M	40	6.8
Type 2N	19	3.2
Type 2 (not specified)	54	9.2
Type 3	17	2.9
Type not determined / unfilled	119	20.3
Total	585	100.0

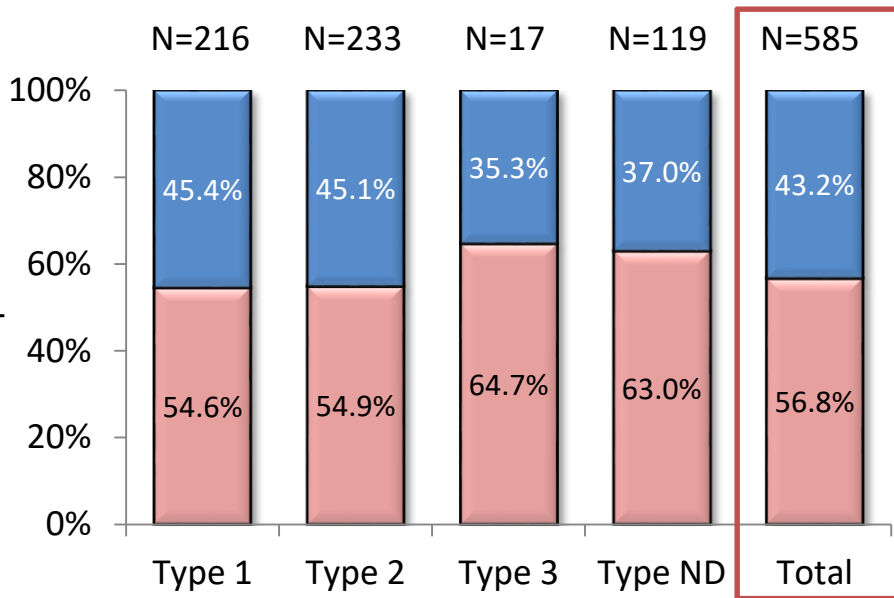
* of them 1 patient with type 1/2N

Sex and current age of patients

N=585

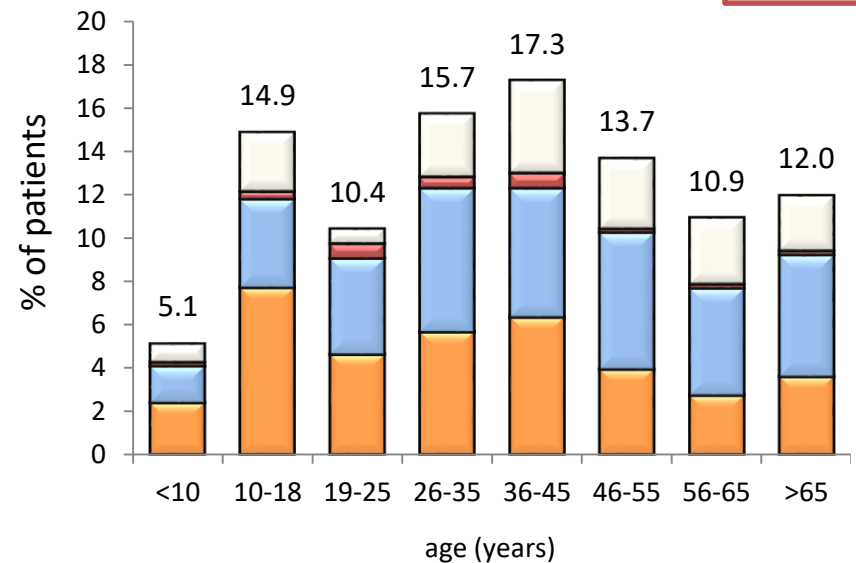
Sex

Men (N=253)
Women (N=332)



Current age*

	Type 1	Type 2	Type 3	Type ND	Total
N	216	233	17	119	585
Mean	34.7	41.2	32.5	42.5	38.8
Median	33.5	41	31	42	39
min - max	2 - 89	1 - 84	1 - 66	4 - 80	1 - 89

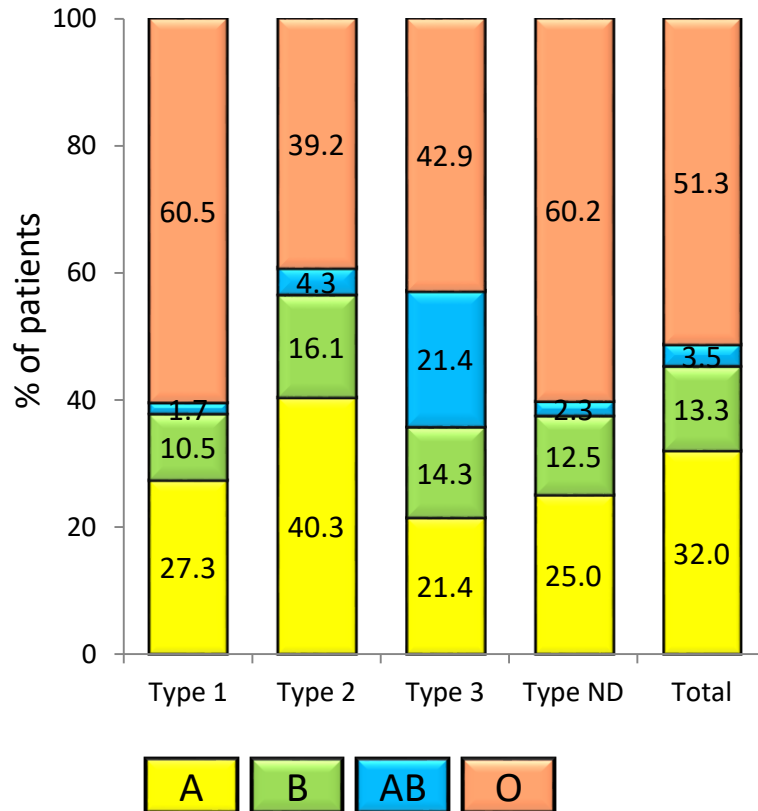


Type ND = not determined or unfilled

* age reached in year 2017

Blood group

N=585

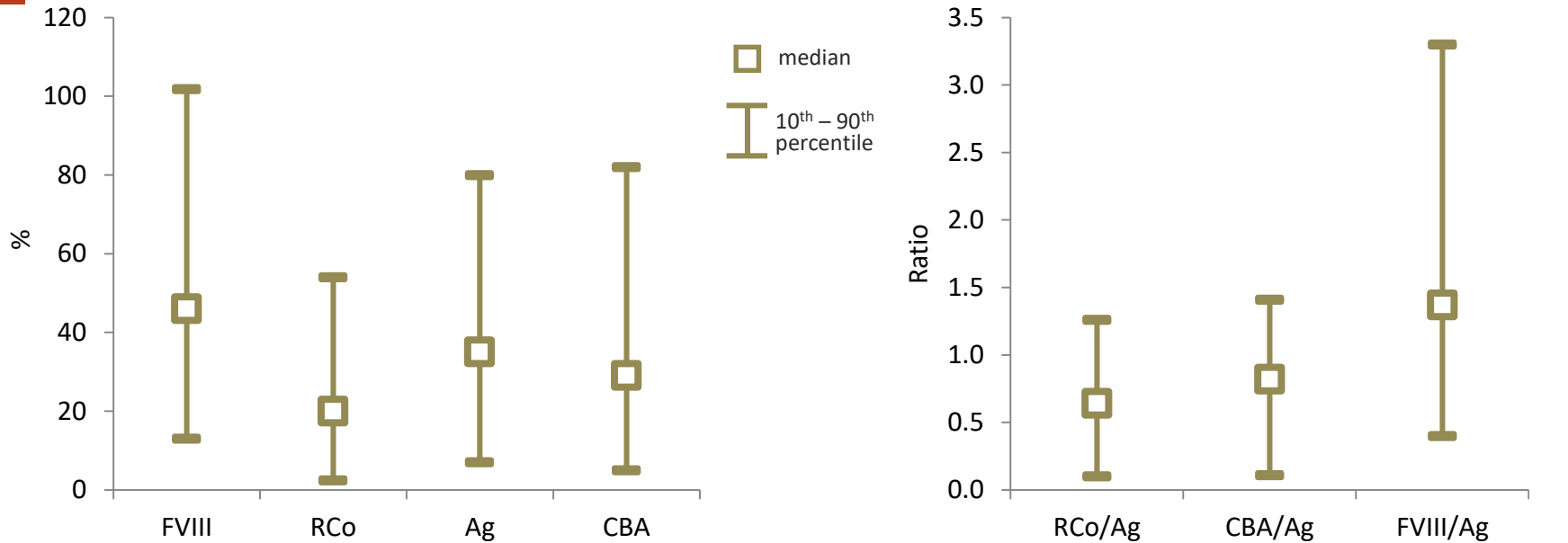


%¹ - % of total N
 %² - % of valid N

Blood group	A	B	AB	O	ND	Valid N	Total N
Type 1	<i>N</i>	47	18	3	104	44	172
	% ¹	21.8	8.3	1.4	48.1	20.4	100.0
	% ²	27.3	10.5	1.7	60.5		100.0
Type 2	<i>N</i>	75	30	8	73	47	186
	% ¹	32.2	12.9	3.4	31.3	20.2	100.0
	% ²	40.3	16.1	4.3	39.2		100.0
Type 3	<i>N</i>	3	2	3	6	3	14
	% ¹	17.6	11.8	17.6	35.3	17.6	100.0
	% ²	21.4	14.3	21.4	42.9		100.0
Type ND	<i>N</i>	22	11	2	53	31	88
	% ¹	18.5	9.2	1.7	44.5	26.1	100.0
	% ²	25.0	12.5	2.3	60.2		100.0
Total	<i>N</i>	147	61	16	236	125	460
	% ¹	25.1	10.4	2.7	40.3	21.4	100.0
	% ²	32.0	13.3	3.5	51.3		100.0

Factor levels

N=585

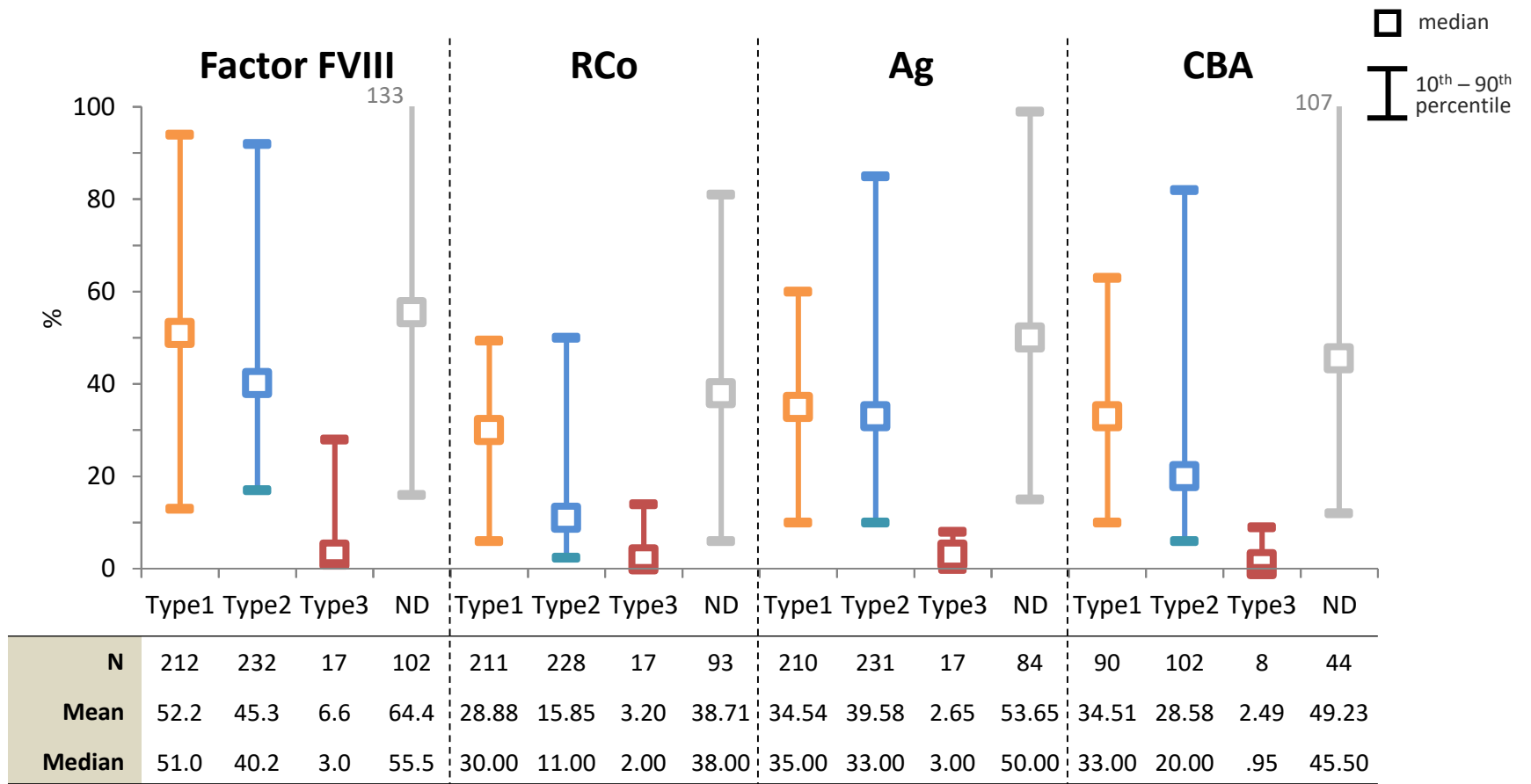


Factor FVIII*	RCo	Ag	CBA		RCo/Ag	CBA/Ag	FVIII/Ag
563	549	542	244	N	532	243	538
50.2	24.3	38.7	33.6	Mean	0.8	0.8	1.9
46 (1–219)	20 (0–135)	35 (0–193)	29 (0–127)	Median (min – max)	0.635 (0–38.03)	0.82 (0–4)	1.365 (0.08–105.63)

* Factor FVIII was assessed by coagulation method in 523 patients, by chromogenic method in 17 patients and method is missing in 45 patients

Factor levels according to type of VWD, part 1

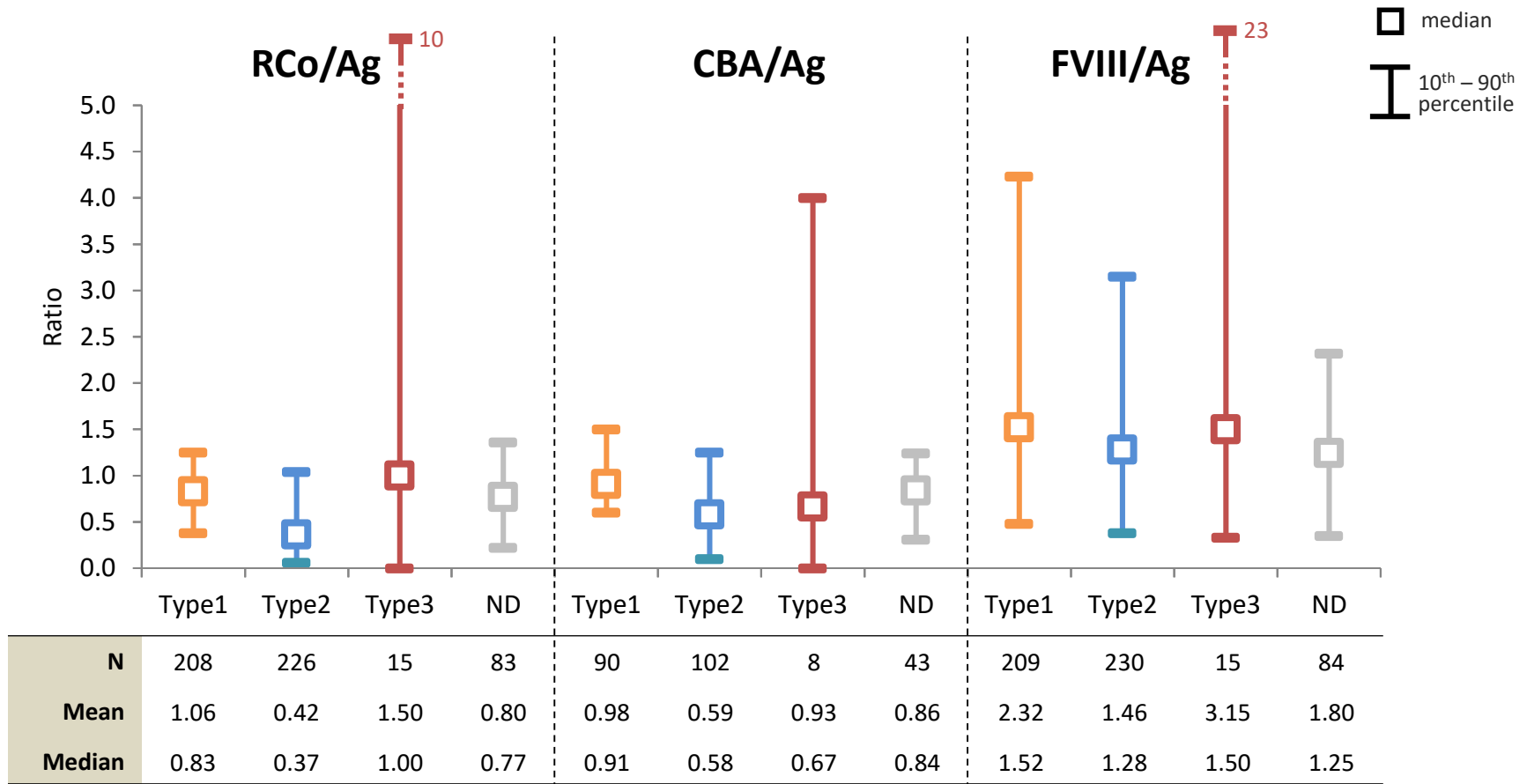
N=585



Type ND = not determined or unfilled

Factor levels according to type of VWD, part 2

N=585

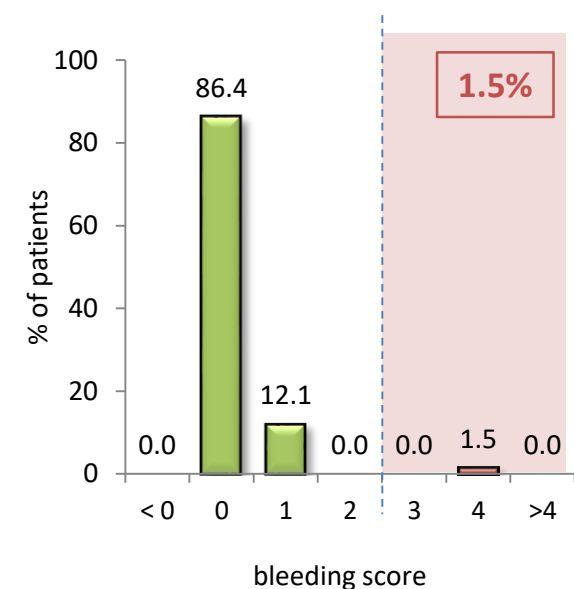
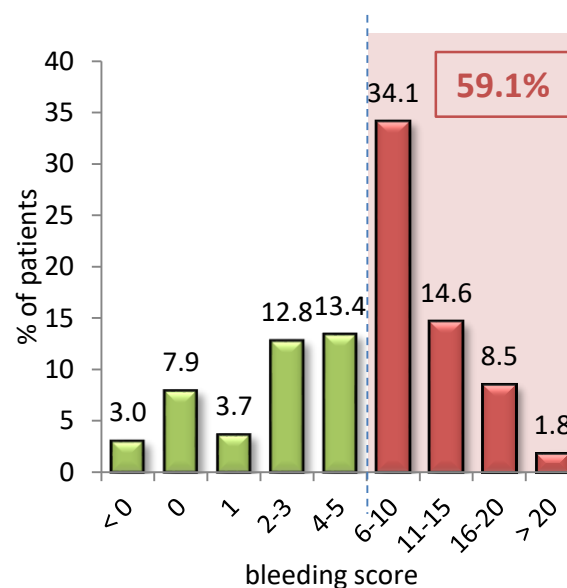
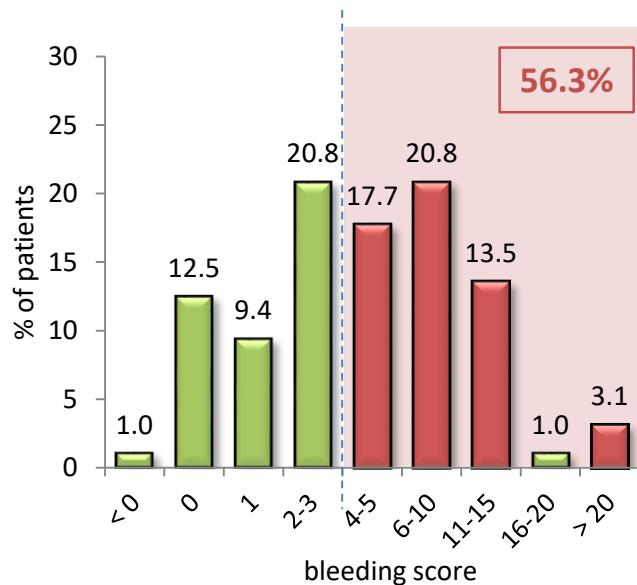


Type ND = not determined or unfilled

Bleeding score¹ according to sex and age

N=326²

	Adult men	Adult women	Children
N	96	164	66
Mean	5.5	7.3	0.2
Median (min - max)	4 (-1 - 25)	6.5 (-3 - 24)	0 (0 - 4)



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

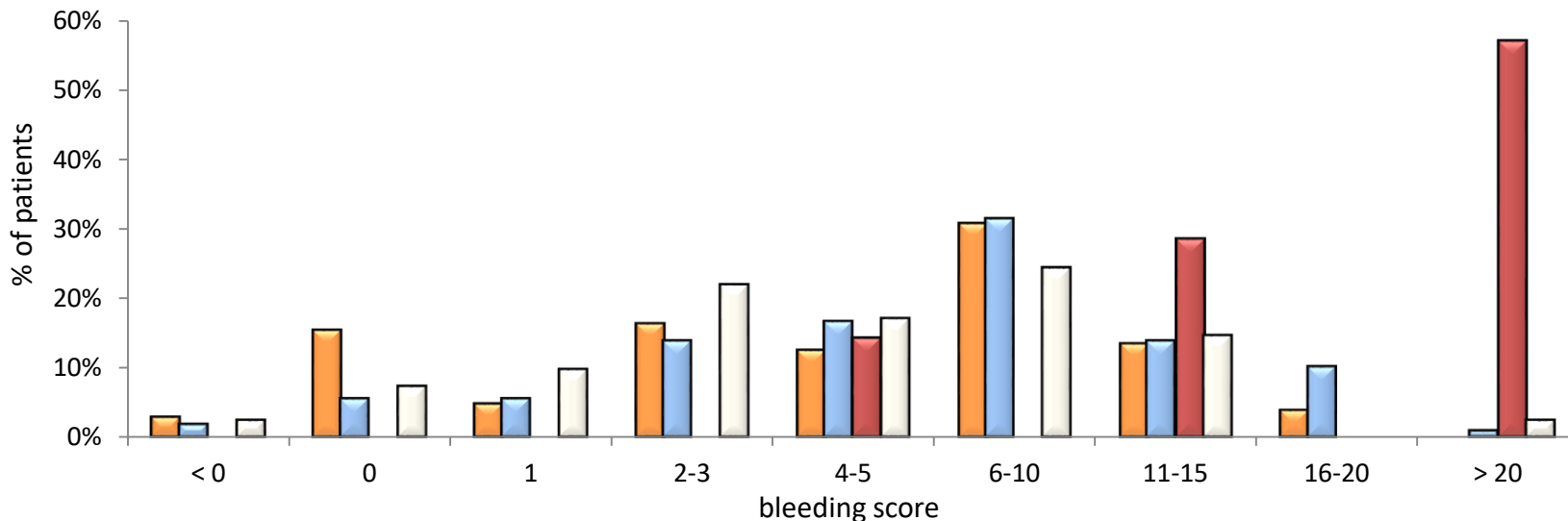
² Missing information on bleeding score in 259 patients.

Bleeding score¹ in adults according to type of disease

N=260²

Bleeding score

	Type 1	Type 2	Type 3	Type ND	Total
N total	157	199	14	98	468
N valid	104	108	7	41	260
Mean	5.6	7.3	17.7	5.4	6.6
Median	5.0	6.0	21.0	4.0	6.0
min - max	-2-18	-1-22	5-25	-3-22	-3-25



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

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

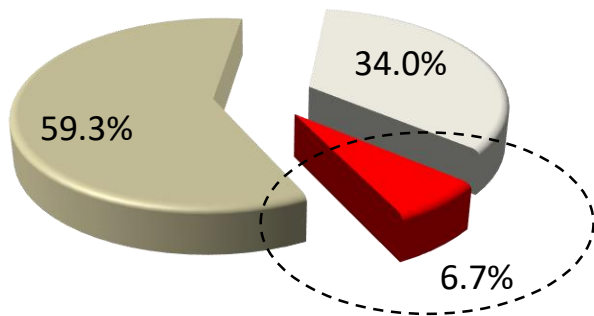
Type ND = not determined or unfilled

Concomitant diseases

N=585

Experienced hepatitis

- Yes (N=39)
- No (N=347)
- Not known (N=199)



24x hepatitis A
7x hepatitis B
4x hepatitis A+B
2x hepatitis C
1x hepatitis A+C

None of the patients is HIV positive.

Annual bleeding rate (ABR) and location of bleeds

N=506¹

	Type 1	Type 2	Type 3	Type ND	Total
ABR					
N valid	198	217	17	74	506
Mean	0.08	0.10	1.06	0.01	0.11
Median	0	0	0	0	0
min – max	0 - 4	0 - 4	0 - 11	0 - 1	0 - 11
SUM	15	21	18	1	55
Location of bleeds					
N_P/N_B²					
Joints	0/-	0/-	2/12	0/-	2/12
Epistaxes	1/1	4/10	0/-	0/-	5/11
Urogenital tract	2/2	2/2	1/4	0/-	5/8
GIT	0/-	3/4	1/1	1/1	5/6
Subcutaneous	0/-	1/1	1/1	0/-	2/2
Muscles	1/1	0/-	0/-	0/-	1/1
Other	2/6	3/3	0/-	0/-	5/9
SUM	6/10	13/20	5/18	1/1	25/49

¹ Missing information on ABR in 79 patients.

Type ND = not determined or unfilled

² N_P = number of patients with bleed; N_B = total number of bleeds treated with substitution of VWF

Consumption of drugs in year 2017

N=585

	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	11 (7/2/1/1)	73 500	6 681.8
<i>of them on prophylaxis*</i>	<i>0 (0/0/0/0)</i>	<i>0</i>	<i>0.0</i>
Haemate P	60 (11/31/6/12)	838 350	13 972.5
<i>of them on prophylaxis*</i>	<i>2 (0/0/2/0)</i>	<i>344 000</i>	<i>172 000.0</i>
Immunate	3 (2/0/1/0)	10 500	3 500.0
<i>of them on prophylaxis*</i>	<i>0 (0/0/0/0)</i>	<i>0</i>	<i>0.0</i>
Wilate	9 (5/3/0/1)	49 000	5 444.4
<i>of them on prophylaxis*</i>	<i>0 (0/0/0/0)</i>	<i>0</i>	<i>0.0</i>
Willfact	1 (0/0/1/0)	196 000	196 000.0
<i>of them on prophylaxis*</i>	<i>1 (0/0/1/0)</i>	<i>196 000</i>	<i>196 000.0</i>
Total	84 (25/36/9/14)	1 167 350	13 897.0
<i>of them on prophylaxis*</i>	<i>3 (0/0/3/0)</i>	<i>540 000</i>	<i>180 000.0</i>
Total - type 1	25	186 500	<u>7 460.0</u>
Total - type 2	36	311 500	<u>8 652.8</u>
Total - type 3	9	604 850	<u>67 205.6</u>
Total - type ND	14	64 500	<u>4 607.1</u>

* Other 3 bleeds were treated with DDAVP

Type ND = not determined or unfilled

Patients on permanent prophylaxis in detail

N=3

No	Centre	Age group	Type of VWD	Type of prophylaxis in 2017	Treatment in 2017	No of applications per week	Total consumption (IU)	ABR in 2017
1	Brno	Child	3	Permanent	Willfact	2	196 000	0
2	Brno	Adult	3	Permanent	Haemate P	2	104 000	0
3	Brno	Adult	3	Permanent	Haemate P	2	240 000	11