

**The status of care for persons with
von Willebrand disease
registered within CNHP registry
Annual Report 2024**

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on behalf of

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

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Persons with Von Willebrand disease



Czech National
Hemophilia
Program

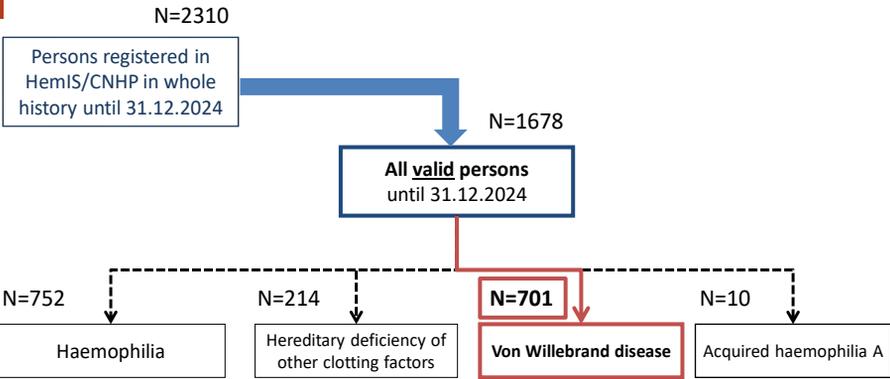


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Sample size



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

Number of patients in participating centres

N=701

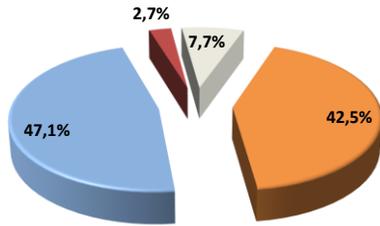
Paediatric centres	Valid patients	
	N	%
Prague – Dpt. of Pediatric Haematology and Oncology, CUH Motol	57	8.1
Ostrava – Dpt. of Pediatric Medicine, UH Ostrava	32	4.6
Pilsen – Pediatric Dpt., UH Pilsen	29	4.1
Brno – Dpt. of Pediatric Haematology, CUH Brno	25	3.6
Olomouc – Dpt. of Pediatric Medicine, UH Olomouc	11	1.6
Ústí n.L. – Pediatric Dpt. – Haematology, Masaryk Hospital	8	1.1
Hradec Králové – Dpt. of Pediatric Medicine, UH HK	6	0.9

Adult centres	Valid patients	
	N	%
Brno – Dpt. Of Clin Hematol, UH Brno	313	44.7
Pilsen – Dpt. of Biochemistry and Hematology, UH Pilsen – UKBH	81	11.6
Ostrava – Blood centre, UH Ostrava	56	8.0
Liberec – Dpt. Of Clin Hematol, Hospital Liberec	34	4.9
Olomouc – Haemato-Oncology Dpt., UH Olomouc	25	3.6
Ústí n.L. – Dpt. Of Clin Hematol, Masaryk Hospital	21	3.0
Nymburk – Center for Thrombosis and Hemostasis, Nymburk Hospital	3	0.4

Centres participating in vWD survey within CNHP registry.

Type of Von Willebrand's disease

N=701



Type of disease	Patients	
	N	%
Type 1*	298	42.5
Type 2	330	47.1
Type 2A	131	18.7
Type 2B	23	3.3
Type 2M	51	7.3
Type 2N	37	5.3
Type 2 (not specified)	88	12.6
Type 3	19	2.7
Type not determined / unfilled	54	7.7
Total	701	100.0

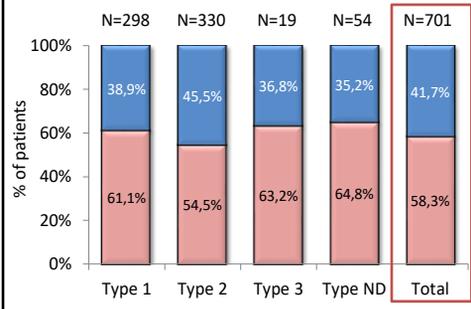
* of them 8 patients with type 1/2N

Sex and current age of patients

N=701

Sex

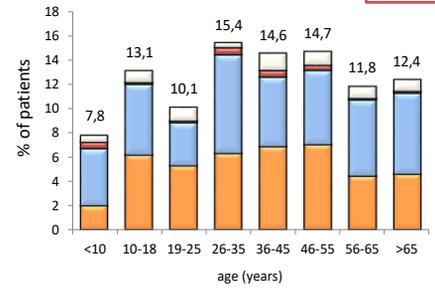
- Men (N=292)
- Women (N=409)



Type ND = not determined or unfilled

Current age*

	Type 1	Type 2	Type 3	Type ND	Total
N	298	330	19	54	701
Mean	38.4	39.2	32.2	40.0	38.8
Median	38	38	31	39.5	38
min - max	4-87	0-88	2-70	3-86	0-88

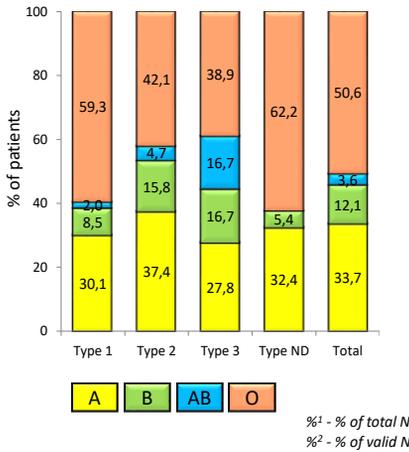


* age reached in year 2024

Median age of Czech persons with vWD is below 40 years.

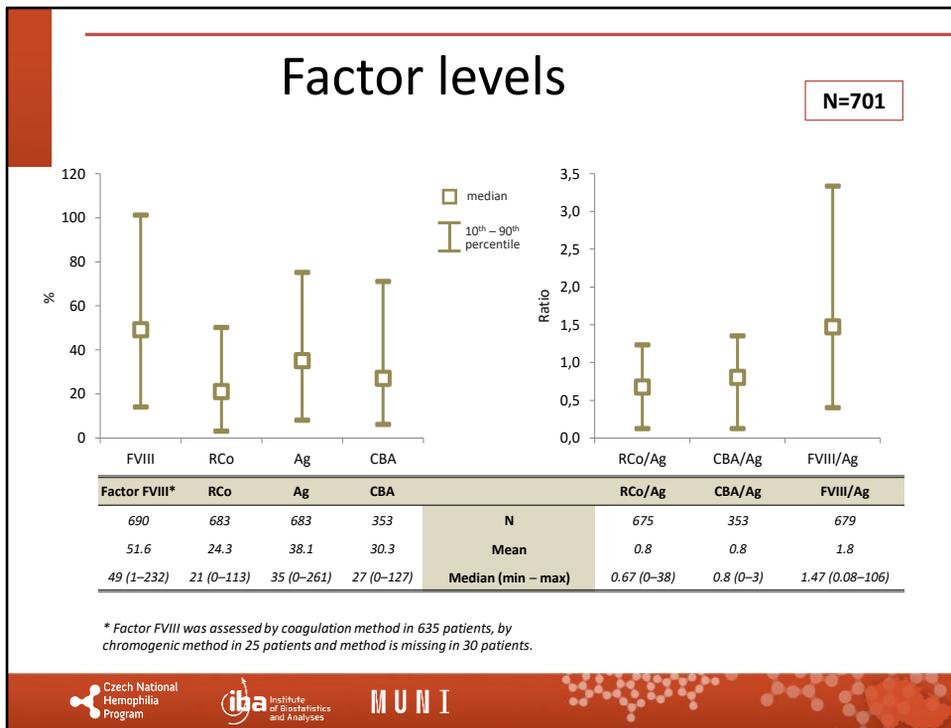
Blood group

N=701



Blood group	A	B	AB	O	ND	Valid N	Total N
Type 1	N	74	21	5	146	52	246
	% ¹	24.8	7.0	1.7	49.0	17.4	100.0
	% ²	30.1	8.5	2.0	59.3	100.0	
Type 2	N	104	44	13	117	52	330
	% ¹	31.5	13.3	3.9	35.5	15.8	100.0
	% ²	37.4	15.8	4.7	42.1	100.0	
Type 3	N	5	3	3	7	1	19
	% ¹	26.3	15.8	15.8	36.8	5.3	100.0
	% ²	27.8	16.7	16.7	38.9	100.0	
Type ND	N	12	2	0	23	17	54
	% ¹	22.2	3.7	0.0	42.6	31.5	100.0
	% ²	32.4	5.4	0.0	62.2	100.0	
Total	N	195	70	21	293	122	701
	% ¹	27.8	10.0	3.0	41.8	17.4	100.0
	% ²	33.7	12.1	3.6	50.6	100.0	

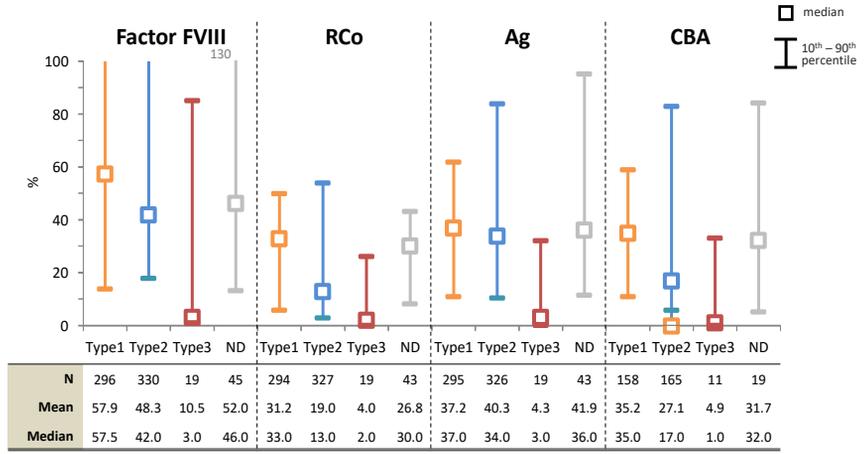
Well known predominance of BG O is confirmed within the registry.



This slide is giving just general overview as it is not related to different vWD subtypes.

Factor levels according to type of VWD, part 1

N=701

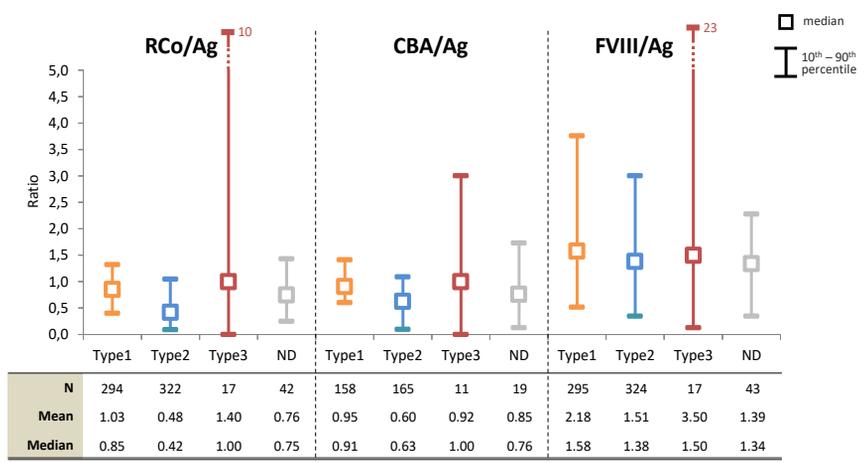


Type ND = not determined or unfilled

This and further slide show factor levels related to respective vWD subtypes. These data correlate well with published findings.

Factor levels according to type of VWD, part 2

N=701



Type ND = not determined or unfilled

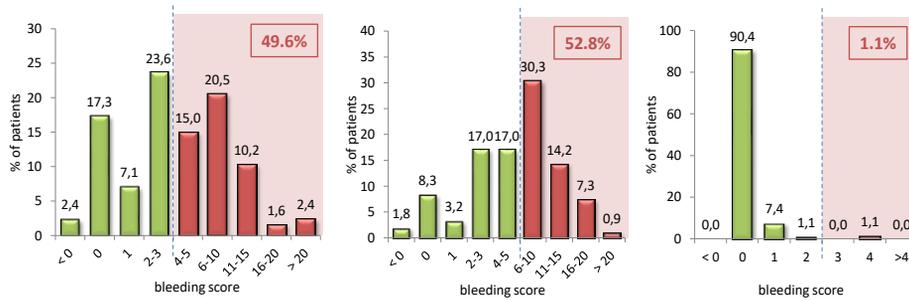


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Bleeding score¹ according to sex and age

N=439²

	Adult men	Adult women	Children
N	127	218	94
Mean	5.0	6.8	0.1
Median (min - max)	3 (-2 - 25)	6 (-2 - 24)	0 (0 - 4)



¹ Adult and Pediatric Vincenza VWD Bleeding Questionnaire and Scoring System

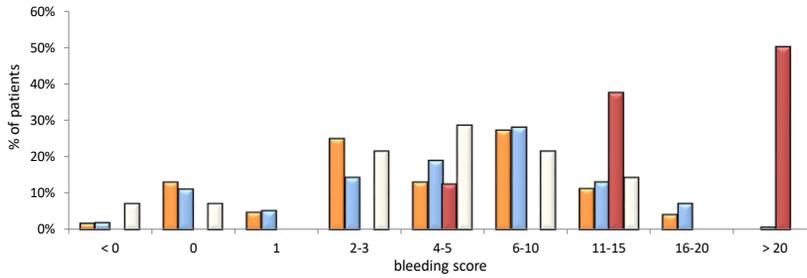
² Missing information on bleeding score in 262 patients.

Children with no previous “bleeding challenges“ are having the lowest bleeding score. In women, almost 53% have bleeding score over 5. (similar since 2020))

Bleeding score¹ in adults according to type of disease

N=345²

Bleeding score	Type 1	Type 2	Type 3	Type ND	Total
N total	241	256	14	43	554
N valid	169	154	8	14	345
Mean	5.4	6.5	17.1	5.5	6.1
Median	5.0	5.0	17.5	4.5	5.0
min - max	-2-20	-1-22	5-25	-2-14	-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

Type 3 has obviously the most heavy bleeding score.

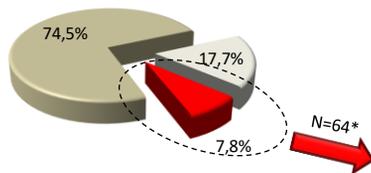
Concomitant diseases

N=701

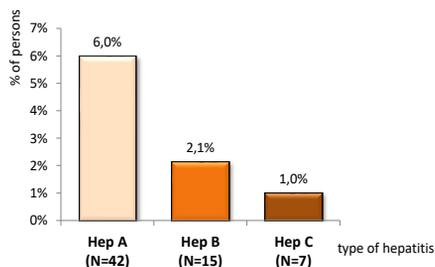
(Ever) Experienced hepatitis

- Yes (N=55)
- No (N=522)
- Not known (N=124)

None of the patients is HIV positive.



*Total of 64 cases of hepatitis in 55 persons. One person may have more types of hepatitis recorded.



Hepatitis and/or HIV prevalence in vWD patients within CNHP registry

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

	Type 1	Type 2	Type 3	Type ND	Total	N=701
ABR						
N valid	298	330	19	54	701	
Mean	0.04	0.12	1.37	0.02	0.12	
Median	0	0	0	0	0	
min-max	0-7	0-16	0-9	0-1	0-16	
SUM	13	41	26	1	81	
Location of bleeds						
N_p/N_B^1						
Joints	1/1	2/4	4/6	0/-	7/11	
Epistaxes	0/-	3/20	4/18	0/-	7/38	
Urogenital tract	-/-	-/-	-/-	-/-	-/-	
GIT	2/3	0/-	0/-	0/-	2/3	
Subcutaneous	0/-	2/6	0/-	0/-	2/6	
Muscles	1/1	3/3	0/-	0/-	4/4	
CNS	-/-	-/-	-/-	-/-	-/-	
Metrorrhagia	1/7	0/-	0/-	0/-	1/7	
Other	0/-	3/4	2/2	1/1	6/7	
SUM	5/12	13/37	10/26	1/1	29/76	

¹ N_p = number of patients with bleed;

N_B = total number of bleeds treated with substitution of VWF

Type ND = not determined or unfilled

Annual bleeding rate and locations of bleeds in vWD patients within the registry. Please note, that not all of the bleeds needed a treatment with vWF concentrate. Median ABR for all types of vWD was 0 in 2024.

Consumption of VWF concentrates in year 2024

N=701

	Number of patients treated with substitution of VWF total (type1/type2/type3/typeND)	Total annual consumption ¹ (IU)	Average annual consumption ¹ per treated patient (IU)
Haemate P	66 (17/38/6/5)	1 949 700	29 540.9
<i>of them on prophylaxis*</i>	5 (0/1/4/0)	1 485 000	297 000.0
Wilate	28 (9/15/4/0)	1 137 350	40 619.6
<i>of them on prophylaxis*</i>	5 (0/1/4/0)	831 350	166 270.0
Fanhdi	1 (1/0/0/0)	5 000	5 000.0
<i>of them on prophylaxis*</i>	0 (0/0/0/0)	0	0.0
Total	95 (27/53/10/5)	3 092 050	32 547.9
<i>of them on prophylaxis*</i>	10 (0/2/8/0)	2 316 350	231 635.0
Total - type 1	27 (27/0/0/0)	201 100	7 448.1
Total - type 2	53 (0/53/0/0)	966 600	18 237.7
Total - type 3	10 (0/0/10/0)	1 887 850	188 785.0
Total - type ND	5 (0/0/0/5)	36 500	7 300.0

* permanent prophylaxis

Type ND = not determined or unfilled

¹ The consumption of Haemate P, Wilate and Fanhdi is given in FVIII units.

Absolute numbers of respective concentrates consumption in this figure refer ONLY to the records within CNHP registry, which have been updated in 2024. The most important information on this slide is „Average annual consumption per treated person“. This aims to reflect nation-wide consumption of factor concentrate per treated patient. Shows well the highest consumption in type 3 vWD. Please note, that the numbers of treated patients are relatively small and do vary significantly between different concentrates. This is, indeed, a source of potential bias. To increase the data validity, we have to increase the number of treated patients recorded within CNHP registry. On the other hand, for vWD treated patients (excluding type 3 patients) the annual consumption per treated patient should reflect the real situation.

Consumption of other drugs in year 2024

N=701

	Number of patients treated with other drugs total (type1/type2/type3/typeND)	Total annual consumption	Average annual consumption per treated patient
DDAVP (µg)	1 (1/0/0/0)	79	79.0
<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

This slide demonstrates the dose of DDAVP used in patients with vWD. However, very little patients were treated with DDAVP and recorded into the CNHP registry in 2024 as nasal DDAVP is still not available in CZ.

Patients on permanent prophylaxis in detail

N=10

No	Centre	Sex	Age group	Type of VWD	Type of prophylaxis in 2024	Treatment in 2024	No of applications per week	Total consumption (IU*)	ABR in 2024
1	Praha	Male	Child	3	Permanent	Wilate	3	186 000	9
2	Ostrava	Male	Child	3	Permanent	Wilate	2	88 250	0
3	Praha	Female	Child	3	Permanent	Wilate	3	77 600	4
4	Praha	Male	Child	2	Permanent	Wilate	2	89 500	16
5	Ostrava	Female	Adult	3	Permanent	Haemate P + jiný	1.42	200 000 IU (+ 2 448 mg)	5
6	Brno	Female	Adult	3	Permanent	Wilate	2.33	390 000	6
7	Brno	Male	Adult	3	Permanent	Haemate P	2.33	368 000	1
8	Brno	Female	Adult	3	Permanent	Haemate P	2.33	346 000	1
9	Brno	Male	Adult	3	Permanent	Haemate P	2	221 000	0
10	Liberec	Male	Adult	2	Permanent	Haemate P	2	350 000	0

* The consumption is given in FVIII units.

This slide shows consumption of factor concentrate in vWD patients on prophylaxis (often permanent). Number of persons with vWD on prophylaxis is low in CZ. Consumption of vWF used prophylactically in vWD patients is similar, to consumption of FVIII for severe PWH on prophylaxis and is dependent on the weight of the respective patient.