

ČNHP

Současný stav

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Koordinátoři ČNHP



Organizační struktura

- Vznikala postupně “zezdola” po dobu 4 let
- Je demokraticky formovaná a odpovídá současným požadavkům center
- Rozhoduje většina (hlasováním), volební období
- Je formálně zakotvená (deklarace)
- Jednotlivá centra nominují zástupce
 - Zvláště děti, zvláště dospělí
- Zástupci center tvoří KR
- KR volí VR



Ta vybere ze svého středu koordinátory

Podpora odborných společností

- Po svém vzniku ČNHP požádal o podporu tyto odborné společnosti
 - ČHS
 - ČSTH
 - ČPS
- Ty souhlasily, což deklarováno podpisy předsedů pod deklarací ČNHP
- ČNHP však není součástí/strukturou žádné odborné společnosti, ale je mezioborovým gremiem zainteresovaných odborníků



Úspěchy ČNHP (ve spolupráci s ČHS)

- Vytvoření formální sítě center a jejich akreditace za účasti odborné veřejnosti a VZP
 - První audit 2011, Re-audit vždy á 2 roky
 - V roce 2014 i audit MZd a ZP
- Úspěšné jednání s NRC/MZd vedoucí k vynětí úhrady léčby hemofilie z DRG a z “paušálů”
- Úzká spolupráce v rámci projektů Evropské komise EUHASS, EUHANET a stím související “PR” české hemofilické péče



Úspěchy ČNHP

- Spolupráce s WFH, EHC a ISTH
 - Od roku 2010 opět dodáváme data pro WFH po přetržce od roku 1996
 - Jubilejní kongres EHC v Praze 2012
 - Spolupráce na QoL dotaznících pro EU stran léčby hemofilie
- Edukační aktivity v rámci OPVK
- Reprezentativní data o léčbě hemofilie a obdobných onemocnění v zemi
- Tvorba standardů péče, publikace



Plány do budoucna...

- Pravidelné audity center obou úrovní ve spolupráci s odbornými společnostmi, MZd, ZP
 - Pokračování certifikace EU center v rámci EUHANET
- Udržení “výkonové” úhrady antihemofilik
- Rozvíjení systémů standardů a doporučení v rámci péče o vrozené krv. choroby
- Edukace/sběr dat a “feed-back”/zlepšování “PR”
- Spolupráce s odbornými společnostmi
- Trvale udržitelný rozvoj (nejen v rámci OPVK)



The current status of care for persons with haemophilia and von Willebrand's disease registered within CNHP registry

Jan Blatný, Petra Ovesná

on behalf of

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

April 2014

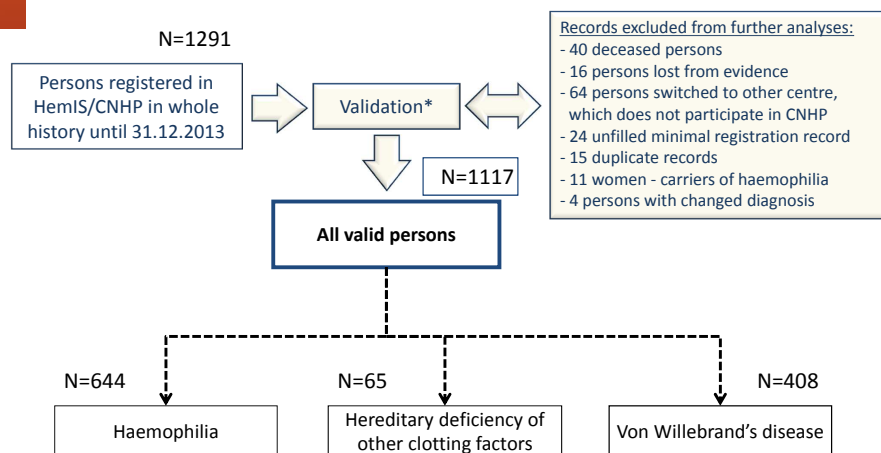


Financování registru ČNHP

- Registr ČNHP vznikl díky projektu OPVK, za přispění Evropského sociálního fondu, MŠMT ČR, Evropské Unie
- V současné době je ČNHP financován z grantu a sponorských darů skrze MU Brno. Sponzoři ani poskytovatelé darů nezasahují do databáze ČNHP, ani jinak neovlivňují sběr a zpracování dat



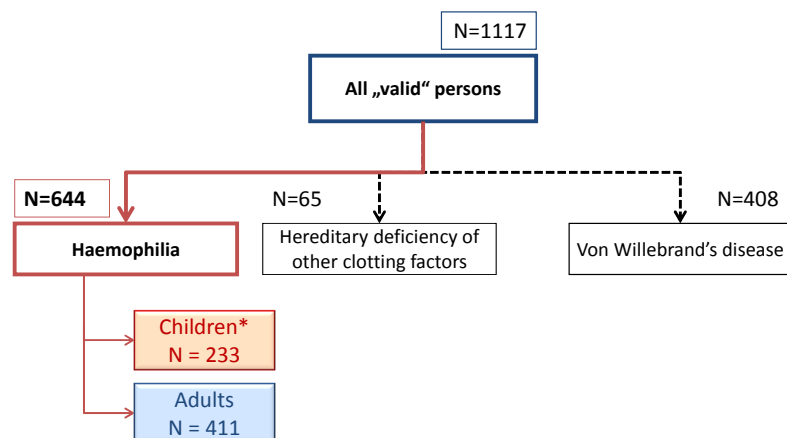
Sample size, valid records



Part A

Persons with haemophilia (PWH)

Sample size



* Persons under 19 years old in 2013

Participating centres in CNHP

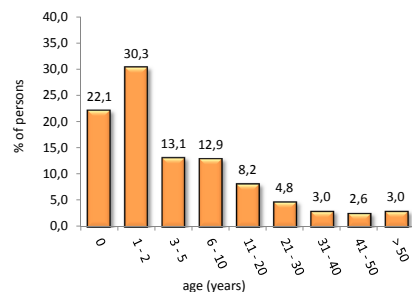
Paediatric centres	Valid persons	
	N	%
FN Motol – Dpt. of Pediatric Haematology and Oncology	78	12.1
FN Brno – DN – Dpt. of Pediatric Haematology	49	7.6
FNHK – Dpt. of Pediatric Medicine	31	4.8
FN Ostrava – Dpt. of Pediatric Medicine	31	4.8
UnL – Pediatric Dpt. – Haematology	30	4.7
CB – Pediatric Dpt.	13	2.0
FN Plzen – Pediatric Dpt.	11	1.7
FN Olomouc – Dpt. of Pediatric Medicine	9	1.4

Adult centres	Valid persons	
	N	%
FN Brno – OKH	132	20.5
FN Ostrava – Blood centre	67	10.4
FN Olomouc – Haemato-Oncology Dpt.	62	9.6
FN Plzen – UKBH	42	6.5
KN Liberec – OKH	34	5.3
CB – OKH	29	4.5
FN a LF HK – IV. IHK	26	4.0

Age

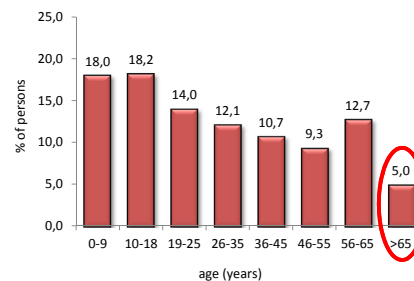
All
N=644

Age at diagnosis (years)	
N	502*
Mean	8.3
Median (min - max)	2 (0 - 81)



* Missing information on year of diagnosis in 142 persons.

Current age (years)	
N	644
Mean	30.6
Median (min - max)	25 (0 - 91)

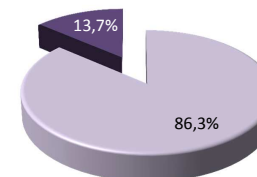


Type and severity of haemophilia I

All
N=644

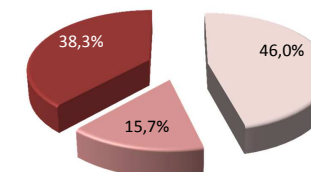
Type of haemophilia

- Haemophilia A (N=556)
- Haemophilia B (N=88)



Severity of haemophilia (N=643*)

- Mild (N=296)
- Moderate (N=101)
- Severe (N=246)



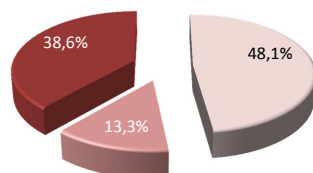
* Severity of haemophilia not known in 1 person.

Type and severity of haemophilia II

All
N=644

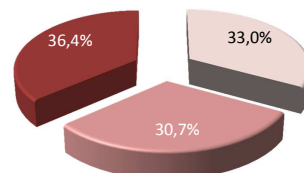
Haemophilia A (N=555¹⁾)

- Mild (N=267)
- Moderate (N=74)
- Severe (N=214)



Haemophilia B (N=88)

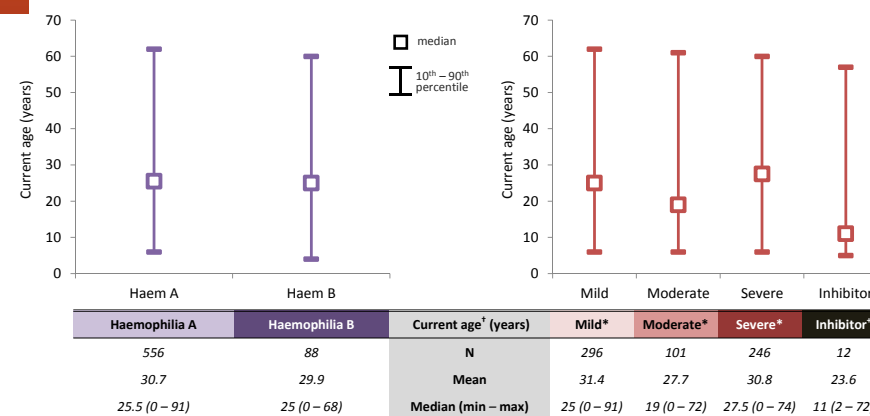
- Mild (N=29)
- Moderate (N=27)
- Severe (N=32)



¹⁾ Severity not known in 1 person with haemophilia A.

Current age according to type and severity of haemophilia

All
N=644



* Current age = age reached in year 2013

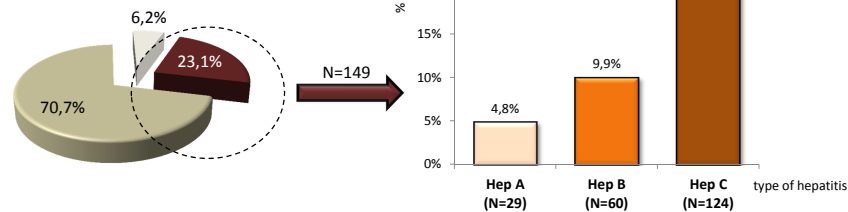
* including persons with inhibitor
* in 2013

Hepatitis experienced

All
N=644

Experienced hepatitis

- Yes (N=149)
- No (N=455)
- Not known (N=40)



Data from last annual report of each person.

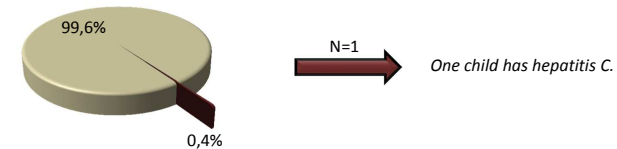
Type of hepatitis not specified in 2 persons. One person may have recorded more types of hepatitis.

Hepatitis experienced

Children
N=233

Experienced hepatitis

- Yes (N=1)
- No (N=232)



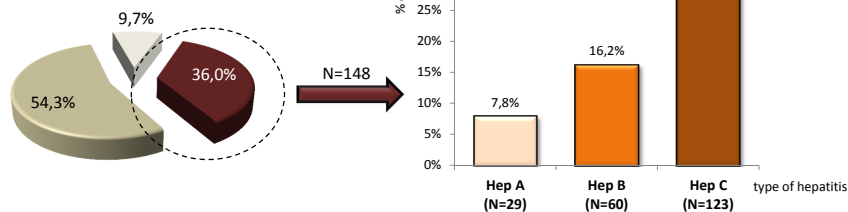
Data from last annual report of each person.

Hepatitis experienced

Adults
N=411

Experienced hepatitis

- Yes (N=148)
- No (N=223)
- Not known (N=40)



Data from last annual report of each person.

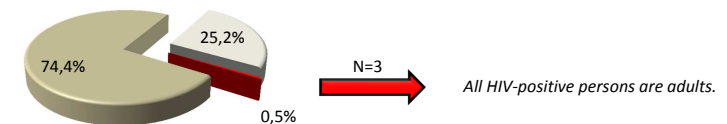
Type of hepatitis not specified in 2 adults. One person may have recorded more types of hepatitis.

HIV

All
N=644

HIV

- Positive (N=3)
- Negative (N=479)
- Not known / not available (N=162)



Data from last annual report of each person.

Data from year 2013 – sample size

	Valid persons			Persons with annual report in 2013			Persons examined in 2013			Persons treated in 2013	
	N	%		N	%		N	%		N	%
All	644	100%	→	634	98.4%	→	533	82.8%	→	370	57.5%
of them with inhibitor				12			12			10	
Children	233	100%	→	229	98.3%	→	213	91.4%	→	139	59.7%
of them with inhibitor				7			7			7	
Adults	411	100%	→	405	98.5%	→	320	77.9%	→	231	56.2%
of them with inhibitor				5			5			3	

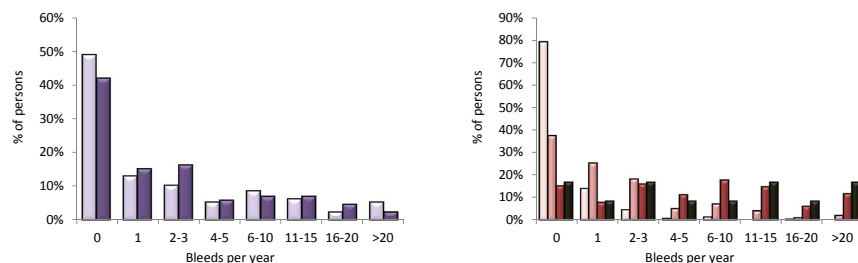
Persons with haemophilia with inhibitors

All
N=634

- inhibitor was recorded in **12 persons** in year 2013
 - 7 children and 5 adults
 - 11 haemophilia A and 1 haemophilia B
 - 10 severe, 1 moderate and 1 mild haemophilia
 - 8 HR and 2 LR (other two patients had in 2013 low level of inhibitor due to by-pass therapy only)
 - 3 patients were treated with rFVIIa, 1 patient with aPCC, other 2 patients both with rFVIIa and aPCC
 - ITT was started in 3 children
 - 2 patients were without any recorded treatment

Frequency of bleeding requiring treatment in 2013

All
N=634



Haemophilia A	Haemophilia B	Frequency of bleeding	Mild*	Moderate*	Severe*	Inhibitor
547	87	N total	286	100	234	12
545	86	N valid	286	99	232	12
4.6	4.0	Mean	0.4	2.7	10 / 6.6†	9.7
1 (0 – 124)	1 (0 – 53)	Median (min – max)	0 (0 – 17)	1 (0 – 30)	5.5 / 4 (0 – 124)	6 (0 – 30)

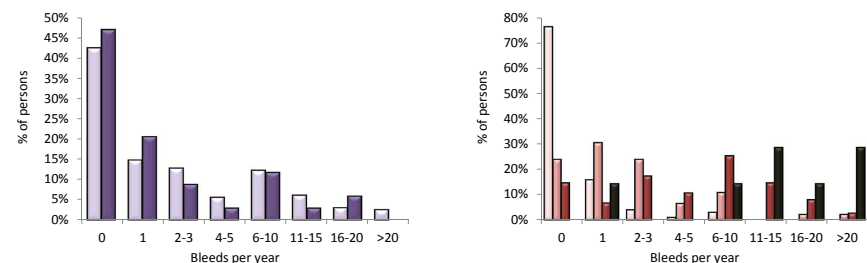
Frequency of bleeding is missing in 207 persons.

* without inhibitor

† mean and median of frequency of bleeding in persons with severe haemophilia without inhibitor on permanent prophylaxis

Frequency of bleeding requiring treatment in 2013

Children
N=229



Haemophilia A	Haemophilia B	Frequency of bleeding	Mild*	Moderate*	Severe*	Inhibitor
195	34	N total	101	46	75	7
195	34	N valid	101	46	75	7
3.9	2.7	Mean	0.5	2.9	7.6 / 7.0†	15.3
1 (0 – 59)	1 (0 – 17)	Median (min – max)	0 (0 – 7)	1 (0 – 22)	6 / 6 (0 – 59)	12 (1 – 30)

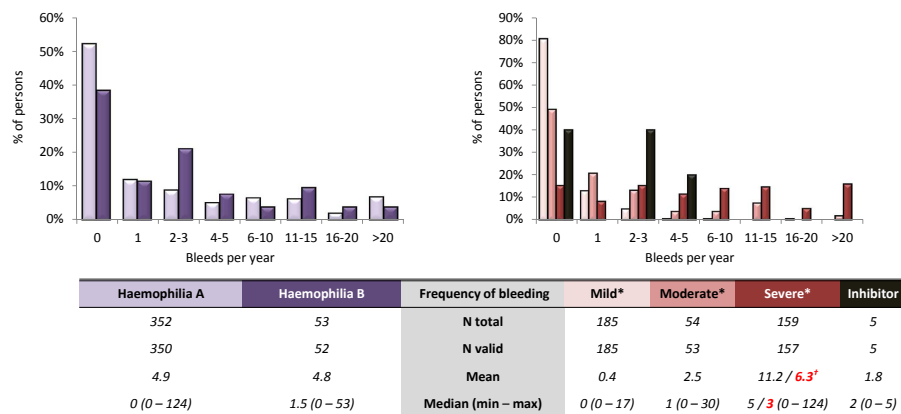
Frequency of bleeding is missing in 19 children.

* without inhibitor

† mean and median of frequency of bleeding in children with severe haemophilia without inhibitor on permanent prophylaxis

Frequency of bleeding requiring treatment in 2013

Adults
N=405



Frequency of bleeding is missing in 188 adults.

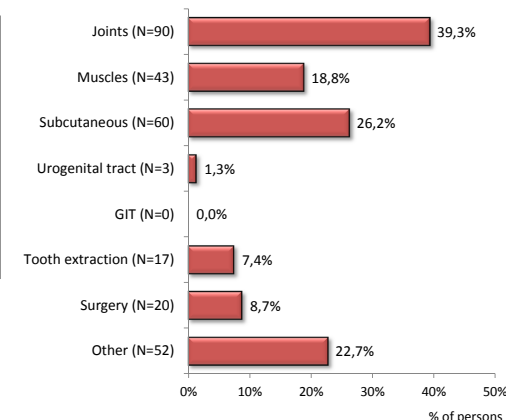
* without inhibitor

† mean and median of frequency of bleeding in adults with severe haemophilia without inhibitor on permanent prophylaxis

Location of (any) bleeds in 2013

Children
N=229

130 (56.8%) children experienced bleeding requiring treatment at least once in year; 856 bleeds were recorded in total, 48 bleeds required hospitalization. 99 (43.2%) children recorded no bleed during year 2013. In total 133 persons recorded bleeds of any location*.

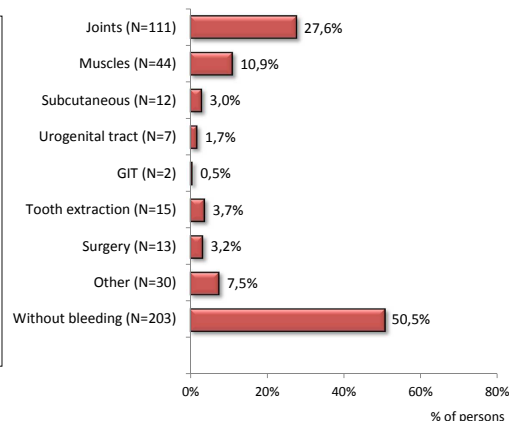


* all recorded bleeds, regardless of requirement of treatment

Location of (any) bleeds in 2013

Adults
N=405

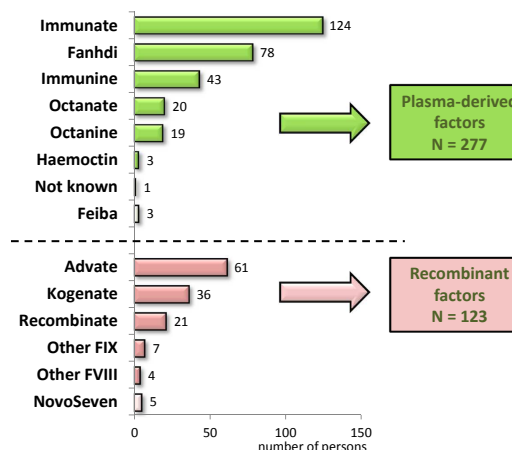
199 (49.5%) adults experienced bleeding requiring treatment at least once in year; 1975 bleeds were recorded in total, 42 bleeds required hospitalization. 203 (50.5%) adults have recorded no bleed during year 2013. Information on frequency of bleeding is missing in 3 examined adults. In total 138 persons recorded bleeds of any location*.



* all recorded bleeds, regardless of requirement of treatment

Treatment

All
N=634



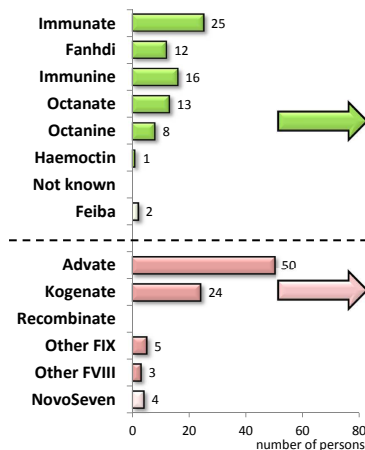
Plasma-derived factors
N = 277

Recombinant factors
N = 123

383 (60.4%) persons received factor concentrates in 2013 (39 of them received more than one type/make of concentrate). Plasma-derived factors were administered more frequently – in 277 (43.7% of all PWHs) persons, whereas recombinant factors in 123 (19.4% of all PWHs) persons. Twenty persons were treated with both plasma-derived and recombinant factor.

Treatment

Children
N=229



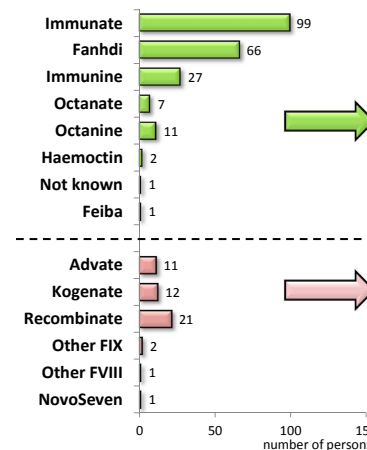
Plasma-derived
factors
N = 72

139 (60.7%) children received factor concentrates in 2013 (22 of them received more than one type/make of concentrate). Plasma-derived factors were administered in 72 (31.4% of all) children, recombinant factors in 77 (33.6% of all) children. Twelve children were treated with both plasma-derived and recombinant factor.

Recombinant
factors
N = 77

Treatment

Adults
N=405



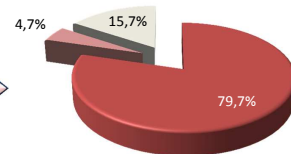
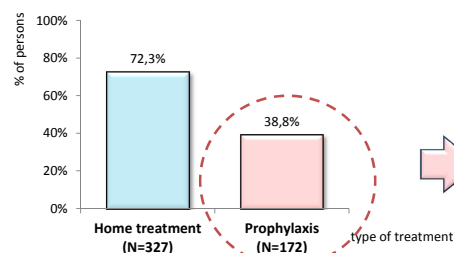
Plasma-derived
factors
N = 205

244 (60.2%) adults received factor concentrates in 2013 (17 of them received more than one type/make of concentrate). Plasma-derived factors were administered more frequently – in 205 (50.6% of all) adults, whereas recombinant factors in 46 (11.4% of all) adults. Eight adults were treated with both plasma-derived and recombinant factor.

Recombinant
factors
N = 46

Type of treatment

All
N=634

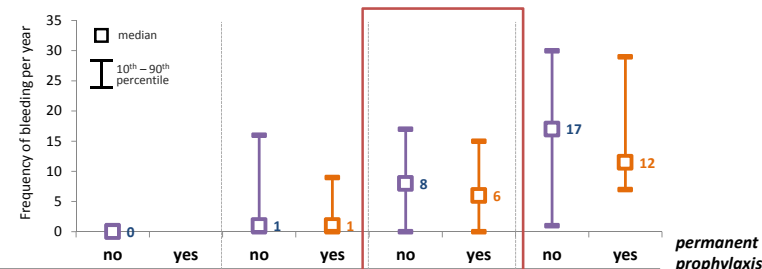


Type of prophylaxis (N=172)

- Permanent (N=137)
- Temporary ≥12 weeks (N=8)
- Temporary <12 weeks (N=27)

Bleeding requiring treatment according to prophylaxis

Children
N=229



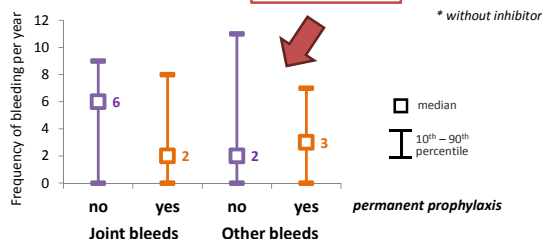
Frequency of bleeding	Mild*		Moderate*		Severe*		Inhibitor	
N	88		38	7	11	62	3	4
Mean	0.5		2.9	3.3	12.3	7.0	16.0	14.8
Median (min – max)	0 (0 – 7)		1 (0 – 22)	1 (0 – 16)	8 (0 – 59)	6 (0 – 45)	17 (1 – 30)	11.5 (7 – 29)
children on permanent prophylaxis	0 (0%)		7 (15.6%)		62 (84.9%)		4 (57.1%)	
% of drugs (FVIII and FIX) consumed by children on permanent prophylaxis	0 %		6.6 %		75.6 %		10.2 %	

* without inhibitor

Joint and other bleeds according to prophylaxis

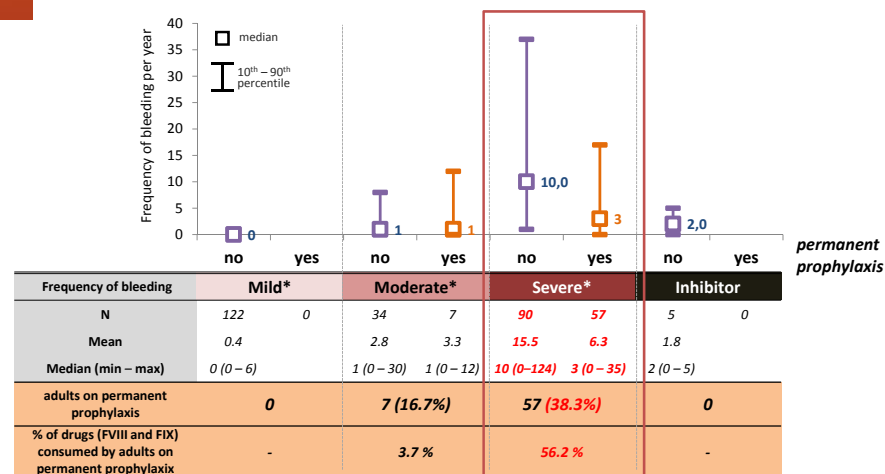
Children
N=229

Frequency of bleeding	Mild*		Moderate*		Severe*		Inhibitor	
N total	88	0	38	7	11	62	3	4
N valid	86	0	38	7	11	61	3	4
JOINT BLEEDS								
Mean	0.2		1.3	1.9	5.7	3.6	9.0	6.3
Median (range)	0 (0-4)		1 (0-10)	1 (0-10)	6 (0-23)	2 (0-29)	9 (0-18)	6 (4-9)
Total bleeding rate	13		50	13	63	222	27	25
OTHER BLEEDS								
Mean	0.4		1.6	1.4	6.5	3.4	7.0	8.5
Median (range)	0 (0-5)		1 (0-12)	0 (0-6)	2 (0-36)	3 (0-16)	8 (1-12)	5.5 (3-20)
Total bleeding rate	33		61	10	72	209	21	34



Bleeding requiring treatment according to prophylaxis

Adults
N=405



Joint and other bleeds according to prophylaxis

Adults
N=405

Frequency of bleeding	Mild*		Moderate*		Severe*		Inhibitor	
N total	122	0	34	7	90	57	5	0
N valid	107	0	25	6	60	35	3	0
JOINT BLEEDS								
Mean	0.1		1.6	3.7	15.2	3.8	0	
Median (range)	0 (0-5)		0 (0-17)	1 (0-12)	10 (0-124)	2 (0-30)	0	
Total bleeding rate	10		41	22	909	133	0	
OTHER BLEEDS								
Mean	0.2		1.1	0.0	2.6	1.6	0.7	
Median (range)	0 (0-3)		0 (0-13)	0	1 (0-24)	1 (0-8)	0 (0-2)	
Total bleeding rate	19		27	0	154	55	2	



Consumption of drugs

All

Drug	Total annual consumption	Number of treated persons	Average annual consumption per treated person	Number of examined persons	Average annual consumption per examined person
FVIII				474	
Immunate	8 447 750 IU	124	68 127.0 IU		17 822.3 IU
Fanhd	5 870 250 IU	78	75 259.6 IU		12 384.5 IU
Octanate	2 080 750 IU	20	104 037.5 IU		4 389.8 IU
Haemoctin	381 500 IU	3	127 166.7 IU		804.9 IU
Advate	4 026 993 IU	61	66 016.3 IU		8 495.8 IU
Kogenate	5 300 750 IU	36	147 243.1 IU		11 183.0 IU
Recombinate	1 858 000 IU	21	88 476.2 IU		3 919.8 IU
Bax 855	96 284 IU	5	19 256.8 IU		203.1 IU
Other recombinant	241 500 IU	4	60 375.0 IU		509.5 IU
FVIII celkem	28 303 777 IU	322	87 899.9 IU		59 712.6 IU
FIX				77	
Immunine	1 633 040 IU	43	37 977.7 IU		21 208.3 IU
Octanine	1 300 880 IU	19	68 467.4 IU		16 894.5 IU
Other recombinant	639 957 IU	4	159 989.3 IU		8 311.1 IU
FIX celkem	3 573 877 IU	58	61 618.6 IU		46 414.0 IU
aPCC					
Feiba	35 000 IU	3	11 666.7 IU		
rFVIIa					
NovoSeven	1140 mg	5	228.0 mg		
Plasma-derived factors - TOTAL*	19 714 170 IU	277	71 170.3 IU		35 585.1 IU
Recombinant factors - TOTAL*	12 163 484 IU	123	98 890.1 IU	554	21 955.7 IU
TOTAL CONSUMPTION *	31 877 654 IU	383	83 231.5 IU		57 540.9 IU

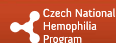
*plasma-derived factors = Immunate, Fanhd, Octanate, Immunine, Octanine, Other plasma-derived
*recombinant factors = Advate, Kogenate, Recombinate, BAX 326, Other recombinant
*TOTAL CONSUMPTION = all mentioned drugs excluding Feiba and NovoSeven

Consumption of drugs

Children

Drug	Total annual consumption	Number of treated persons	Average annual consumption per treated child	Number of examined children	Average annual consumption per examined child
<i>Immunate</i>	1 633 500 IU	25	65 340.0 IU		8 926.2 IU
<i>Fanhdi</i>	1 158 250 IU	12	96 520.8 IU		6 329.2 IU
<i>Octanate</i>	1 692 750 IU	13	130 211.5 IU		9 250.0 IU
<i>Haemoctin</i>	165 000 IU	1	165 000.0 IU		901.6 IU
FVIII				183	
<i>Advate</i>	2 838 993 IU	50	56 779.9 IU		15 513.6 IU
<i>Kogenate</i>	2 200 750 IU	24	91 697.9 IU		12 026.0 IU
<i>BAX 855</i>	65 284 IU	3	21 761.3 IU		356.7 IU
<i>Stude gena 13</i>	241 500 IU	4	60 375.0 IU		1 319.7 IU
<i>FVIII celkem</i>	9 996 027 IU	116	86 172.6 IU		54 623.1 IU
FIX				33	
<i>Immunine</i>	575 140 IU	16	35 946.3 IU		17 428.5 IU
<i>Octanine</i>	267 580 IU	8	33 447.5 IU		8 108.5 IU
<i>Other recombinant</i>	212 214 IU	3	70 738.0 IU		6 430.7 IU
<i>FIX celkem</i>	1 054 934 IU	21	50 235.0 IU		31 967.7 IU
aPCC					
<i>Feiba</i>	23 000 IU	2	11 500.0 IU		
rFVIIa					
<i>NovoSeven</i>	695 mg	4	173.8 mg		
Plasma-derived factors - TOTAL*	5 492 220 IU	72	76 280.8 IU		25 426.9 IU
Recombinant factors - TOTAL*	5 558 741 IU	77	72 191.4 IU	216	25 734.9 IU
TOTAL CONSUMPTION *	11 050 961 IU	139	79 503.3 IU		51 161.9 IU

*plasma-derived factors = Immunate, Fanhdi, Octanate, Immunine, Octanine, Other plasma-derived
 *recombinant factors = Advate, Kogenate, Recombinate, BAX 326, Other recombinant
 *TOTAL CONSUMPTION = all mentioned drugs excluding Feiba and NovoSeven

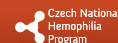


Consumption of drugs

Adults

Drug	Total annual consumption	Number of treated persons	Average annual consumption per treated person	Number of examined adults	Average annual consumption per examined adult
<i>Immunate</i>	6 814 250 IU	99	68 830.8 IU		23 416.7 IU
<i>Fanhdi</i>	4 712 000 IU	66	71 393.9 IU		16 192.4 IU
<i>Octanate</i>	388 000 IU	7	55 428.6 IU		1 333.3 IU
<i>Haemoctin</i>	216 500 IU	2	108 250.0 IU		744.0 IU
FVIII				291	
<i>Advate</i>	1 188 000 IU	11	108 000.0 IU		4 082.5 IU
<i>Kogenate</i>	3 100 000 IU	12	258 333.3 IU		10 652.9 IU
<i>Recombinate</i>	1 858 000 IU	21	88 476.2 IU		6 384.9 IU
<i>BAX 855</i>	31 000 IU	2	15 500.0 IU		106.5 IU
<i>FVIII celkem</i>	18 307 750 IU	206	88 872.6 IU		62 913.2 IU
FIX				44	
<i>Immunine</i>	1 057 900 IU	27	39 181.5 IU		24 043.2 IU
<i>Octanine</i>	1 033 000 IU	11	93 936.4 IU		23 484.1 IU
<i>Other recombinant</i>	427 743 IU	1	427 743.0 IU		9 721.4 IU
<i>FIX celkem</i>	2 518 943 IU	37	68 079.5 IU		57 248.7 IU
aPCC					
<i>Feiba</i>	12 000 IU	1	12 000.0 IU		
rFVIIa					
<i>NovoSeven</i>	445 mg	1	445.0 mg		
Plasma-derived factors - TOTAL*	14 221 950 IU	205	69 375.4 IU		42 453.6 IU
Recombinant factors - TOTAL*	6 604 743 IU	46	143 581.4 IU	335	19 715.7 IU
TOTAL CONSUMPTION *	20 826 693 IU	244	85 355.3 IU		62 169.2 IU

*plasma-derived factors = Immunate, Fanhdi, Octanate, Immunine, Octanine, Other plasma-derived
 *recombinant factors = Advate, Kogenate, Recombinate, BAX 326, Other recombinant
 *TOTAL CONSUMPTION = all mentioned drugs excluding Feiba and NovoSeven



Děkuji za pozornost

- Poděkování lékařům, sestrám a datamanagerům všech center ČNHP
- Poděkování IBA
- Poděkování sponsorům projektu
- Více na www.cnhp.cz

