The status of care for persons with haemophilia registered within CNHP registry
Annual Report 2022

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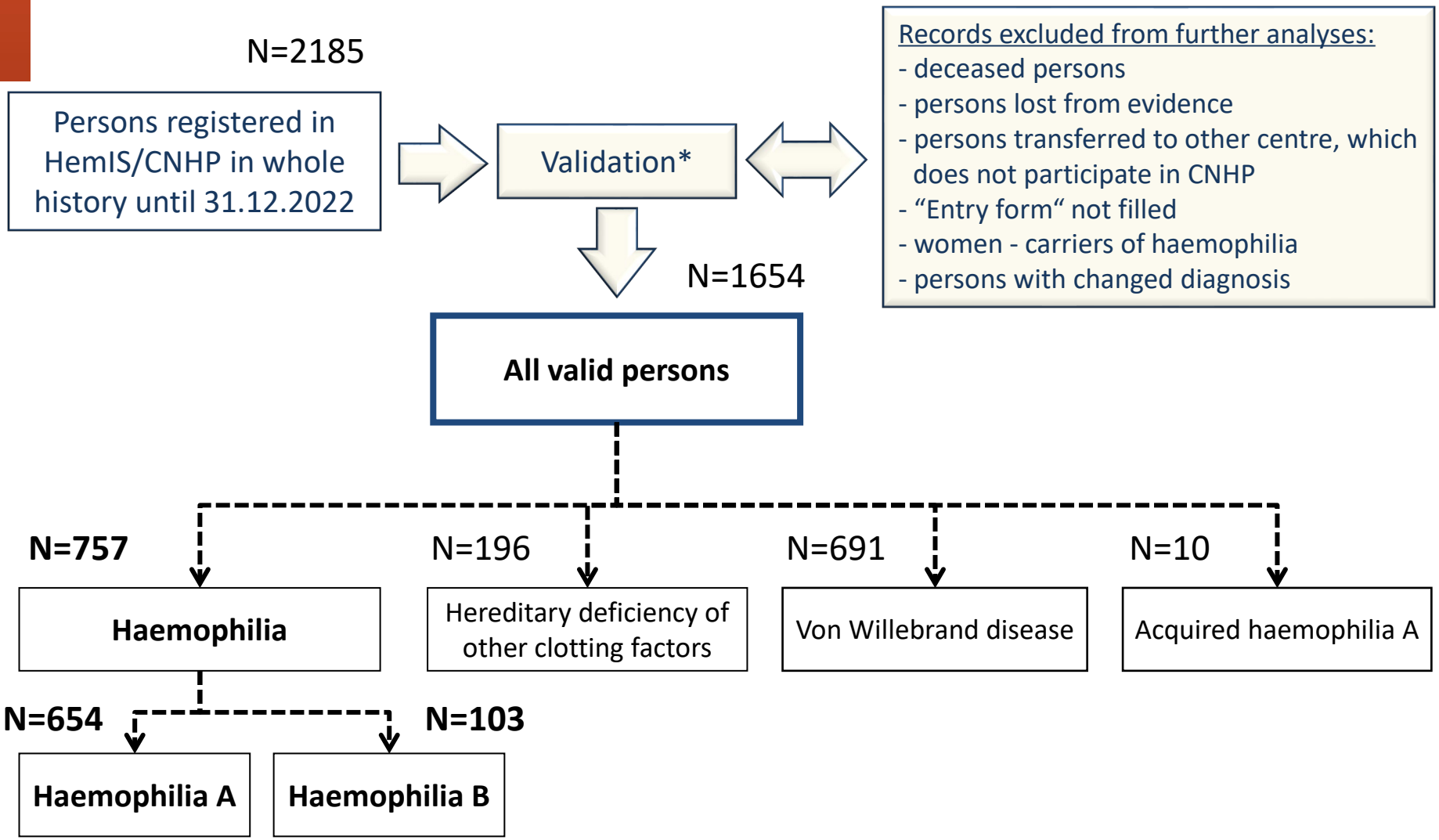
on behalf of

Centres contributing to CNHP registry

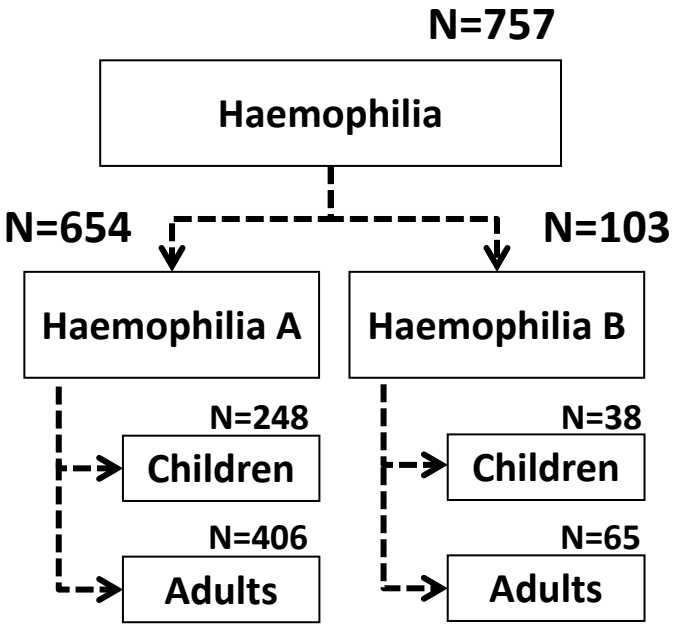
(Czech National Haemophilia Programme)

Export date: March 28, 2023

Sample size, valid records



Persons with haemophilia (PWH)



Centres participating in CNHP

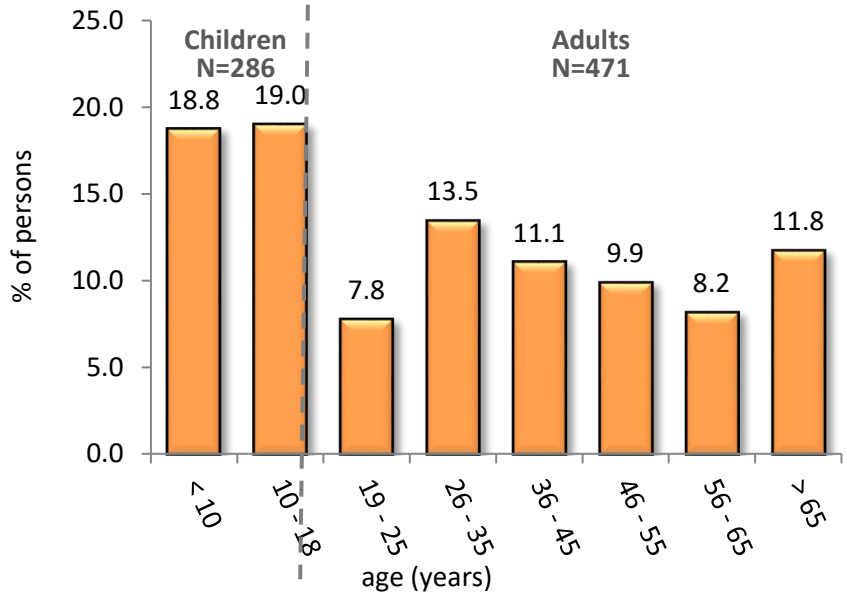
Paediatric centres	Valid persons N	%
Prague – Dpt. of Pediatric Haematology and Oncology, CUH Motol	107	14.1
Brno – Dpt. of Pediatric Haematology, CUH Brno	64	8.5
Hradec Králové – Dpt. of Pediatric Medicine, UH HK	32	4.2
Ostrava – Dpt. of Pediatric Medicine, UH Ostrava	24	3.2
Olomouc – Dpt. of Pediatric Medicine, UH Olomouc	19	2.5
Ústí n.L. – Pediatric Dpt. – Haematology, Masaryk Hospital	19	2.5
České Budejovice – Pediatric Dpt., Hospital CB	18	2.4
Pilsen – Pediatric Dpt., UH Pilsen	13	1.7

Adult centres	Valid persons N	%
Brno – Dpt. Of Clin Hematol, UH Brno	170	22.5
Ostrava – Blood centre, UH Ostrava	77	10.2
Olomouc – Haemato-Oncology Dpt., UH Olomouc	60	7.9
Pilsen – Dpt. of Biochemistry and Hematology, UH Pilsen	52	6.9
Liberec – Dpt. Of Clin Hematol, Hospital Liberec	46	6.1
Ústí n.L. – Dpt. Of Clin Hematol, Masaryk Hospital	29	3.8
České Budějovice – Dpt. Of Clin Hematol, Hospital CB	27	3.6

Basic demographics

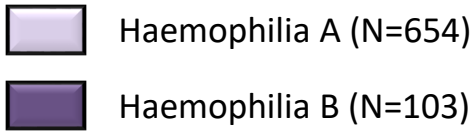
All
N=757

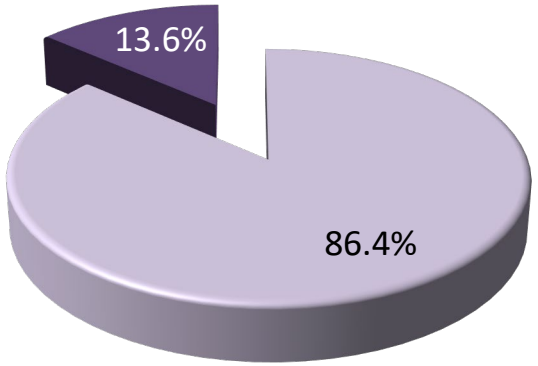
	Actual age* (years)
N	*757*
Mean	*32.3*
Median (min - max)	*30 (0 – 90)*



** age reached in year 2022*

Type of haemophilia





Three children with haemophilia were born in 2022.

All
N=757

Persons with haemophilia and inhibitors in 2022

Active inhibitors were recorded in <u>24 persons</u> in the end of year 2022

- 5 inhibitors in children with severe HA and 1 inhibitor in children with mild HA newly developed in 2022

PWH with inhibitors:

- 17 children and 7 adults
- 23 haemophilia A and 1 haemophilia B
- 20 in severe, 2 in moderate and 2 in mild haemophilia
- 18 high-titre and 6 low-titre (<5BU)
- 13 high response and 7 low response inhibitors; this information not available in 4 PWH with inhibitors
- 21 patients were treated with emicizumab
  - 13 patients were treated only with emi, 7 patients with emi and rFVIII, and 1 patient with emi, FVIII and rFVIIa during the year
- 1 patient with haemophilia B was treated only with rFVIIa

ITT:

- One patient has already been on-going ITT in 2022 (started earlier, died in 2022).
- Two patients (with newly developed inhibitors) started ITT in 2022.

ABR and treatment regimens in patients with inhibitor

N=24

	Type	Year of birth	Severity	ITT	Emi	By-pass	Titre	Responder	ABR	Joint / other
1	HA	2021	Severe		Yes	OD	high	HR	1	0 / 2
2	HA	2021	Severe		Yes		low	LR	1	0 / 1
3	HA	2021	Severe		Yes		low	LR	2	0 / 2
4	HA	2021	Severe	Yes	Yes		low	LR	3	1 / 2
5	HA	2020	Mild		Yes		high	HR	2	0 / 2
6	HA	2020	Severe	Yes	Yes		high	LR	0	0 / 1
7	HA	2019	Severe		Yes		low	*NA*	0	0 / 0
8	HA	2018	Severe		Yes		low	LR	0	0 / 0
9	HA	2018	Severe		Yes		high	*NA*	0	0 / 0
10	HA	2017	Severe		Yes		high	HR	0	0 / 0
11	HA	2016	Severe		Yes		high	*NA*	0	0 / 0
12	HA	2016	Severe		Yes		high	*NA*	0	0 / 0
13	HA	2015	Severe		Yes		high	HR	0	0 / 0
14	HA	2014	Severe		Yes		high	HR	0	0 / 0
15	HA	2011	Moderate		Yes		high	HR	1	/
16	HA	2004	Severe		Yes		high	HR	0	0 / 0
17	HA	1977	Severe		Yes		high	HR	0	0 / 0
18	HA	1975	Severe		Yes		high	HR	0	0 / 0
19	HA	1971	Severe		Yes		high	HR	0	0 / 0
20	HA	1971	Severe				high	LR	1	1 / 0
21	HA	1956	Severe		Yes		high	HR	0	0 / 0
22	HA	1941	Moderate		Yes		high	HR	0	0 / 0
23	HA	1941	Mild	Yes			low	LR	1	0 / 1
24	HB	2007	Severe			Permanent px	high	HR	11	8 / 3

new in 2022

NA not available

N=24

ABR according to treatment regimen in PWH with inhibitor

Diagnosis	ITT	Emi/by-pass prophylaxis	N	ABR (mean)	ABR (median, min-max)	Joint / other bleeds (median)
Haemophilia A	Yes	Emi px	2	1.50	1.5 (0-3)	0.5 / 1
		OD	1	1.00	1 (1-1)	0 / 1
	No	Emi px	19	0.37	0 (0-2)	0 / 0
		OD	1	1.00	1 (1-1)	1 / 0
Haemophilia B	No	BPA permanent	1	11.00	11 (11-11)	8 / 3

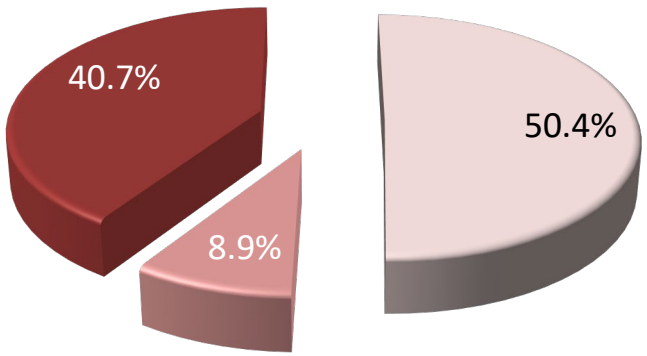
Demographic characteristics Haemophilia A

Severity of haemophilia A

Haem A
N=653*

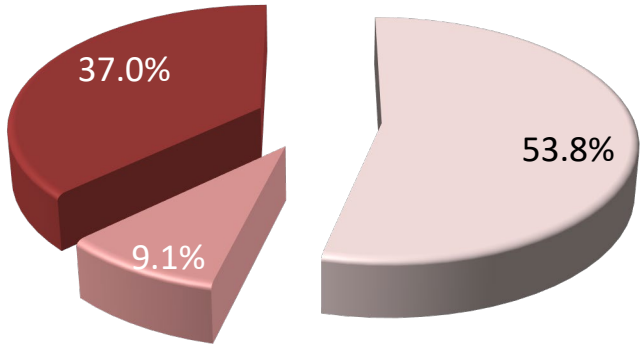
Children (N=248)

- Mild (N=125)
- Moderate (N=22)
- Severe (N=101)



Adults (N=405*)

- Mild (N=218)
- Moderate (N=37)
- Severe (N=150)

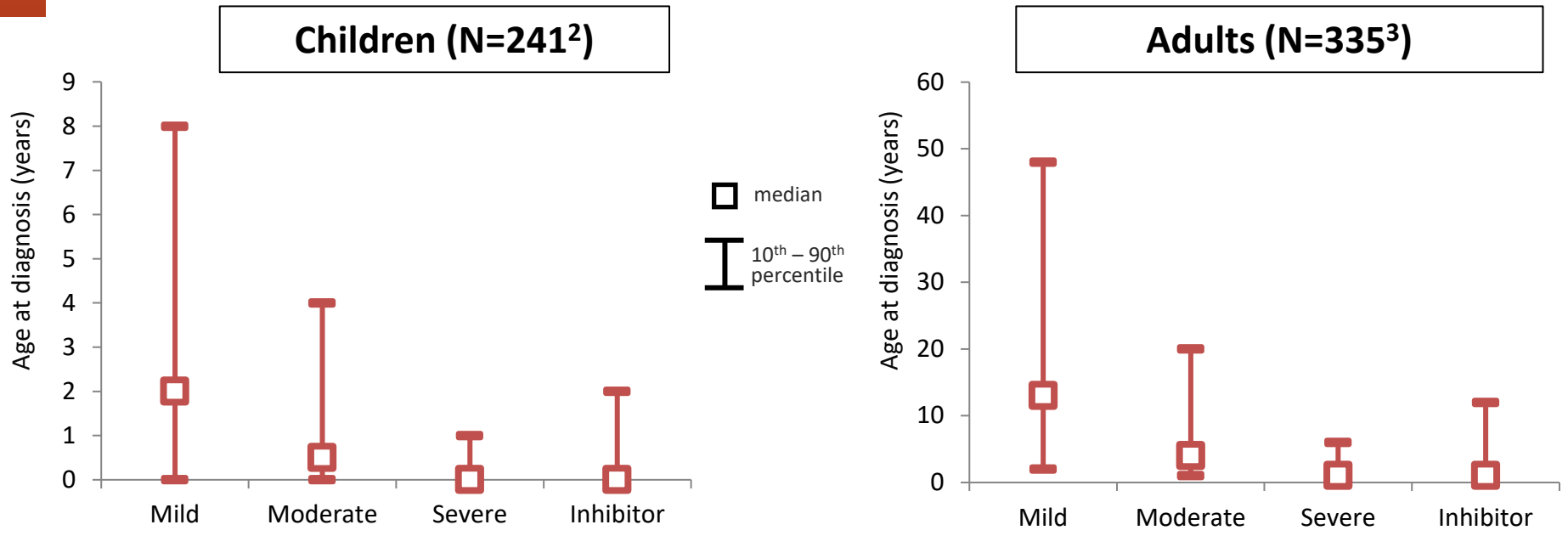


** Severity of haemophilia not known in 1 adult with haemophilia A.*

Age at diagnosis according to severity of haemophilia A

Haem A
N=653[1]

[1] *severity of haemophilia not known in 1 adult*



Mild*	Moderate*	Severe*	Inhibitor+	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor+
122	*22*	*97*	*16*	**N valid**	*191*	*31*	*113*	*7*
3.0	*1.6*	*0.6*	*0.6*	**Mean**	*19.8*	*7.9*	*2.3*	*3.7*
2 (0 – 18)	*0.5 (0 – 10)*	*0 (0 – 7)*	*0 (0 – 4)*	**Median (min – max)**	*13 (0 – 68)*	*4 (0 – 32)*	*1 (0 – 38)*	*1 (0 – 12)*

[2] *Missing information on year of diagnosis in 7 children.*

[3] *Missing information on year of diagnosis in 70 adults.*

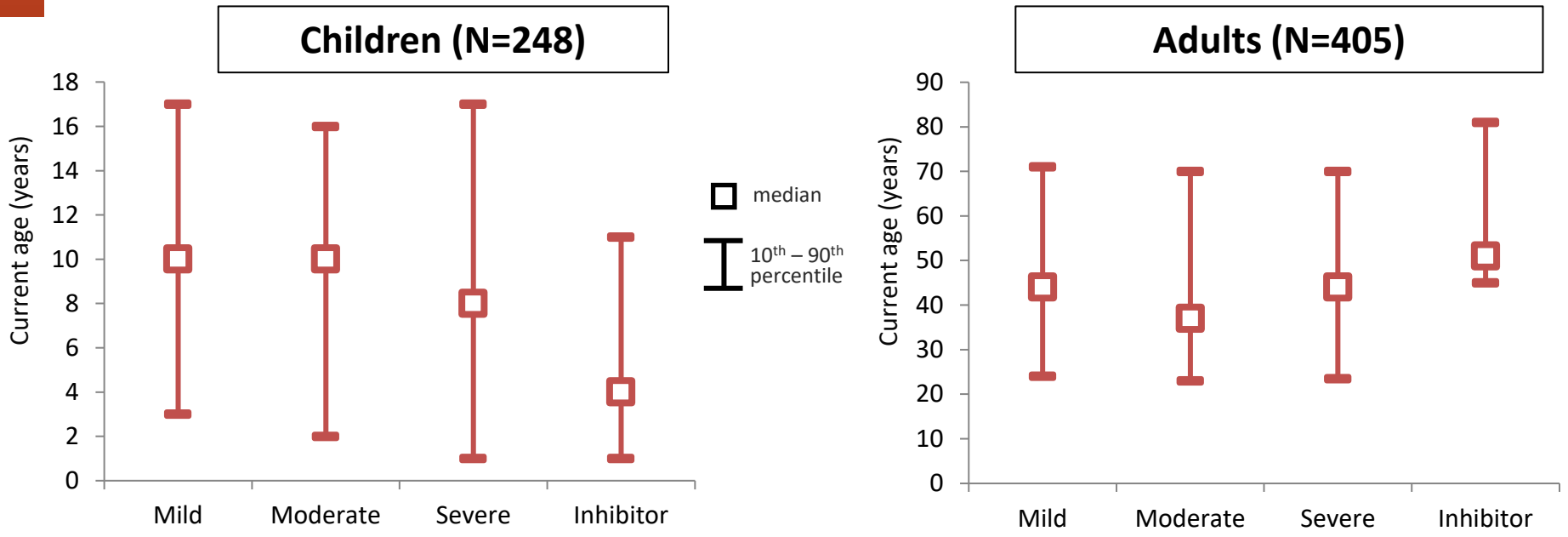
** including persons with inhibitor*

+ in 2022

Actual age according to severity of haemophilia A

Haem A
N=653[1]

[1] *severity of haemophilia not known in 1 adult*



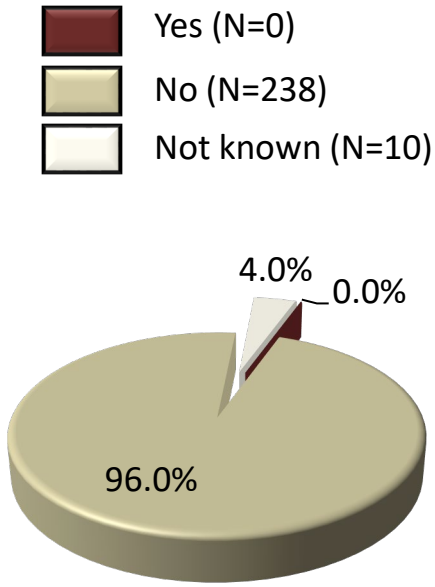
Mild*	Moderate*	Severe*	Inhibitor+	Current age++ (years)	Mild*	Moderate*	Severe*	Inhibitor+
125	*22*	*101*	*16*	**N valid**	*218*	*37*	*150*	*7*
10.0	*9.8*	*8.8*	*5.0*	**Mean**	*46.4*	*42.6*	*45.6*	*60.3*
10 (0 – 18)	*10 (1 – 17)*	*8 (0 – 18)*	*4 (1 – 18)*	**Median (min – max)**	*44 (19 – 90)*	*37 (19 – 81)*	*44 (19 – 83)*	*51 (45 – 81)*

** including persons with inhibitor*
+ in 2022
++ age reached in year 2022

Hepatitis (ever) experienced

Children
Haem A
N=248

Experienced hepatitis



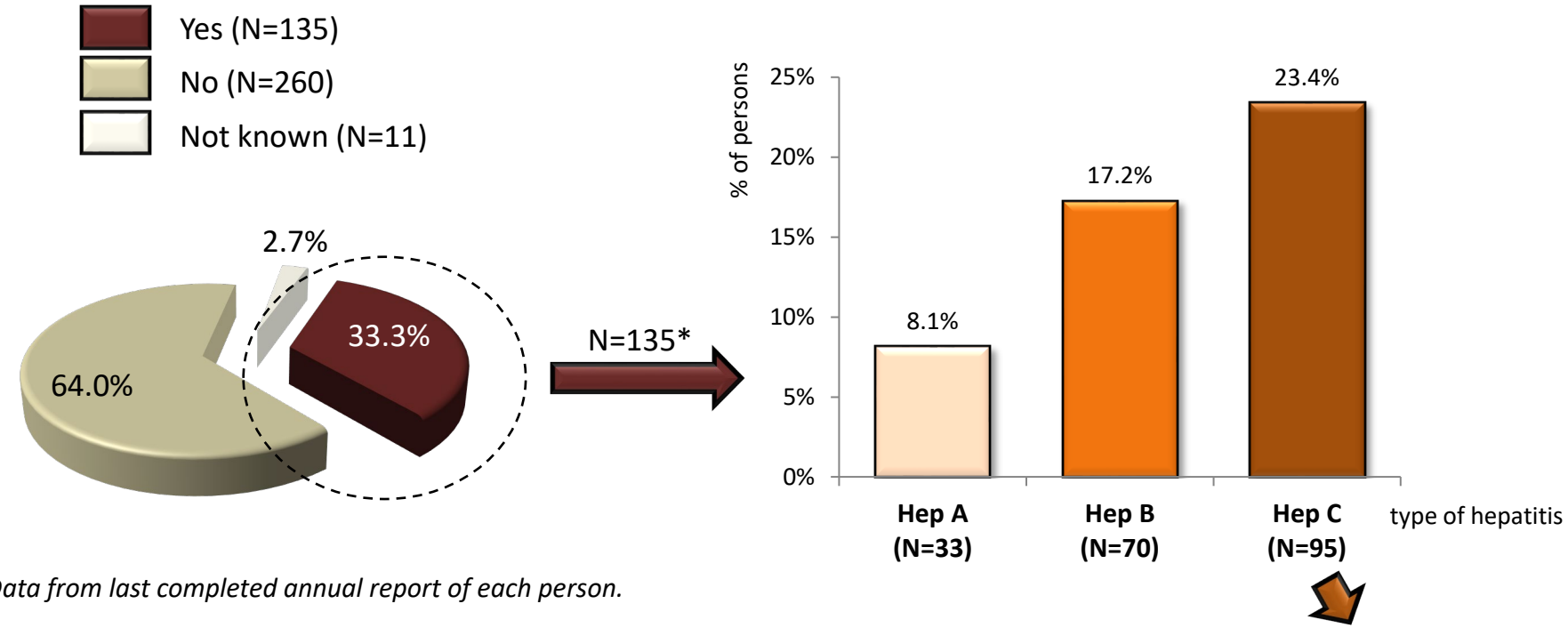
No child has hepatitis.

Data from last completed annual report of each person.

Hepatitis (ever) experienced

Adults
Haem A
N=406

Experienced hepatitis



Data from last completed annual report of each person.

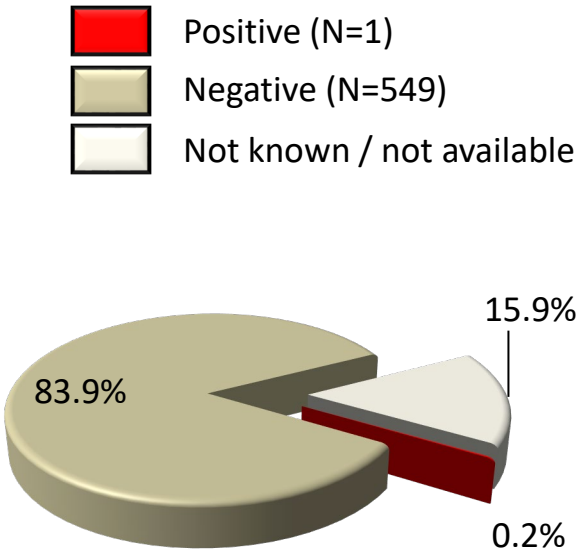
**Total of 198 cases of hepatitis in 135 persons. One person may have more types of hepatitis recorded.*

19 adults are HCV RNA positive

HIV

All
Haem A
N=654

HIV



N=1 (+ 1 in another centre)

All HIV-positive persons are adults.

Data from last completed annual report of each person.

Treatment outcomes and bleeding frequency Haemophilia A

All
Haem A
N=654

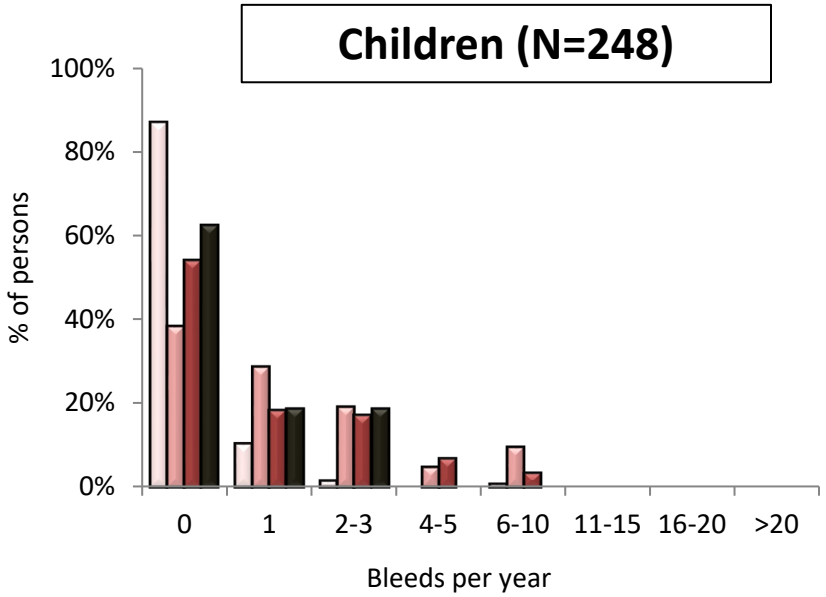
Data from year 2022 – sample size

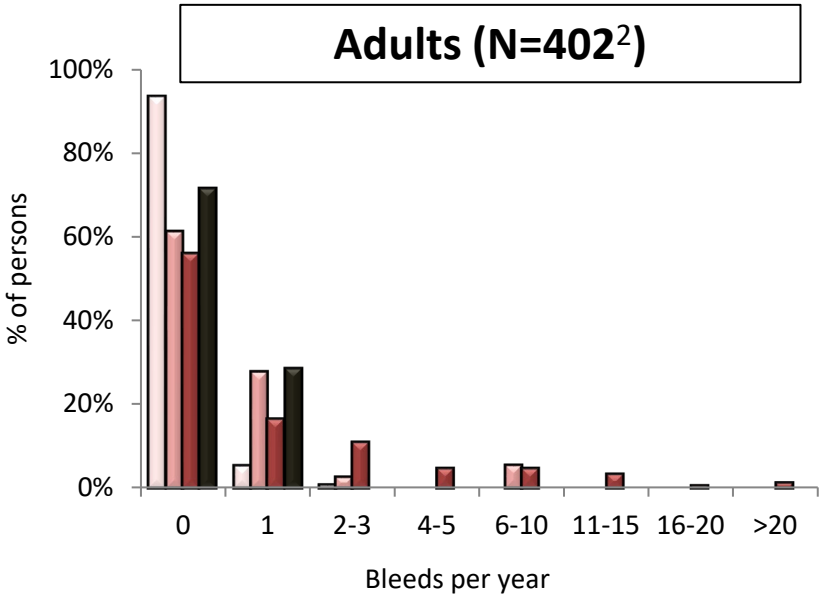
	Valid persons			Persons with valid annual report			Persons examined			Persons treated	
	N	%		N	%		N	%		N	%
All	654	100%	→	644	98.5%	→	479	73.2%	→	336	51.4%
of them with inhibitor	23			23			23			23	
Children	248	100%	→	248	100.0%	→	215	86.7%	→	135	54.4%
of them with inhibitor	16			16			16			16	
Adults	406	100%	→	396	97.5%	→	264	65.0%	→	201	49.5%
of them with inhibitor	7			7			7			7	

Frequency of bleeding requiring treatment in 2022

Haem A
N=653[1]

[1] *severity of haemophilia not known in 1 adult*





Mild*	Moderate*	Severe*	Inhibitor	Frequency of bleeding	Mild*	Moderate*	Severe*	Inhibitor
124	*21*	*87*	*16*	**N valid**	*217*	*35*	*143*	*7*
0.2	*1.9*	*1.1*	*0.6*	**Mean**	*0.1*	*0.7*	*2.0*	*0.3*
0 (0 – 7)	*1 (0 – 10)*	*0 (0 – 7)*	*0 (0 – 3)*	**Median (min – max)**	*0 (0 – 3)*	*0 (0 – 6)*	*0 (0 – 35)*	*0 (0 – 1)*
108 (87.1%)	*8 (38.1%)*	*47 (54%)*	*10 (62.5%)*	**N (%) with no bleed**	*203 (93.5%)*	*22 (61.1%)*	*81 (55.9%)*	*5 (71.4%)*

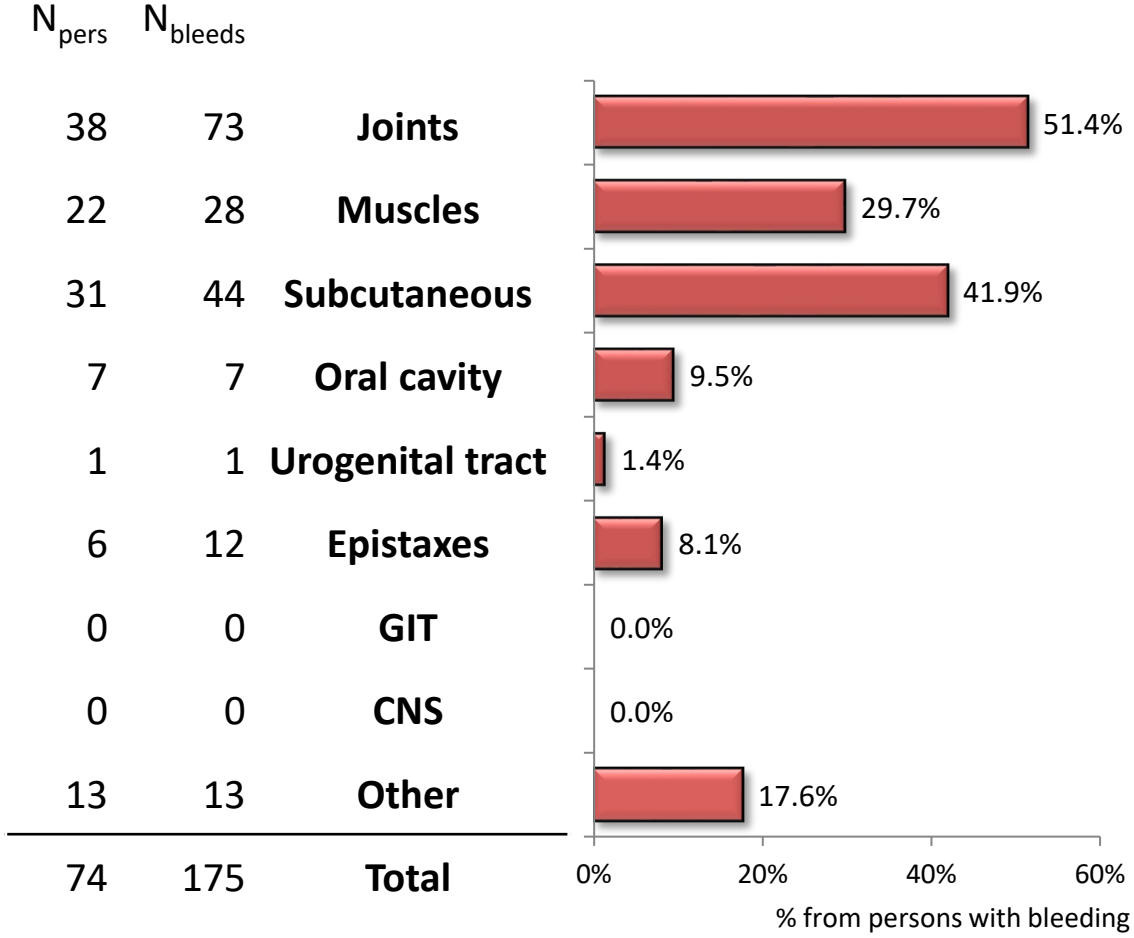
** without inhibitor*

[2] *Frequency of bleeding is missing in 3 adults.*

Location of bleeds in 2022

Children
Haem A
N=248

75 (30.2%) children experienced bleeding at least once in year; 178 bleeds were recorded in total, 12 bleeds required hospitalization.
74 of these 75 children have recorded location of their bleeds. Localization is not known in 1 child.
173 (69.8%) children recorded no bleed during year 2022.

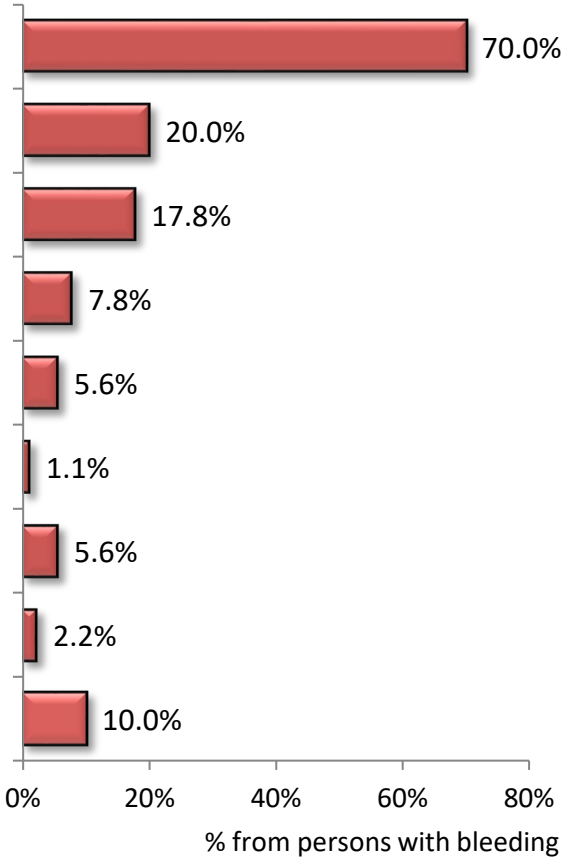


Location of bleeds in 2022

Adults
Haem A
N=403[1]

91 (22.6%) adults experienced bleeding at least once in year; 338 bleeds were recorded in total, 17 bleeds required hospitalization.
90 of these 91 adults have recorded location of their bleeds. Localization is not known in 1 adult.
312 (77.4%) adults have recorded no bleed during year 2022.

N_{pers}	N_{bleeds}		
63	247	**Joints**	70.0%
18	32	**Muscles**	20.0%
16	21	**Subcutaneous**	17.8%
7	11	**Oral cavity**	7.8%
5	5	**Urogenital tract**	5.6%
1	1	**Epistaxes**	1.1%
5	7	**GIT**	5.6%
2	2	**CNS**	2.2%
9	11	**Other**	10.0%
90	337	**Total**	

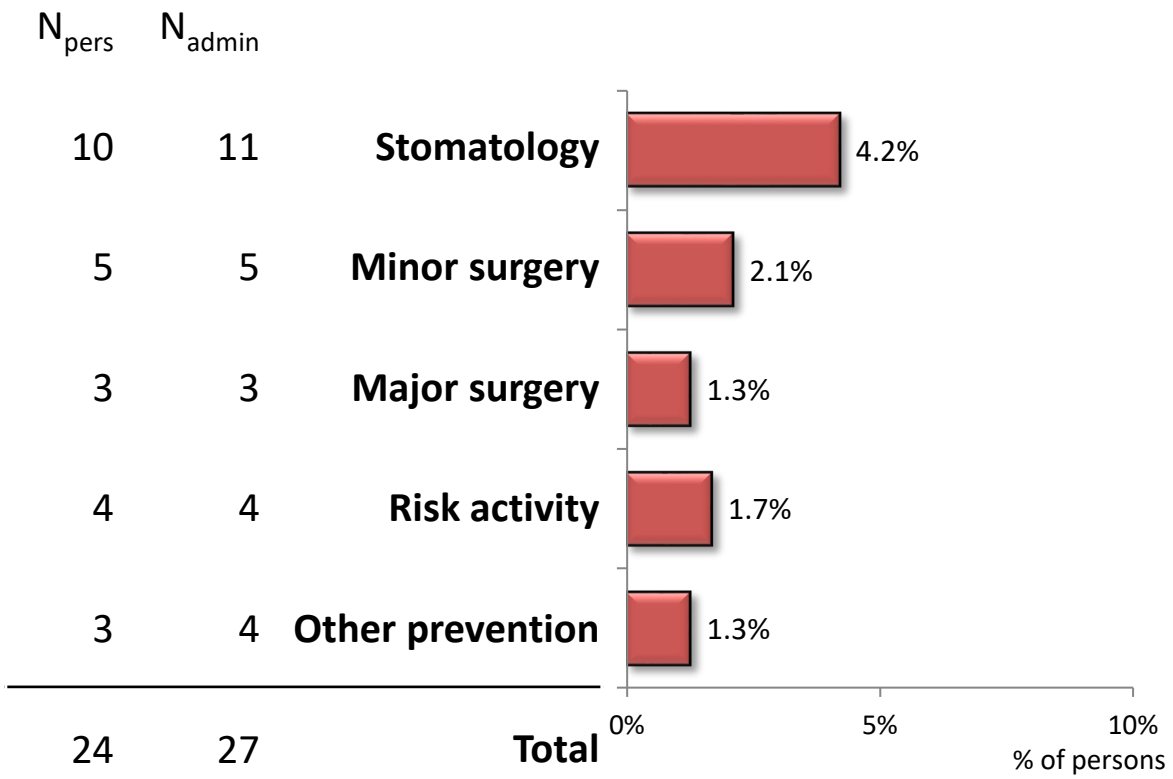


[1]*Frequency of bleeding is missing in 3 adults.*

Preventive administration in 2022

Children
Haem A
N=248

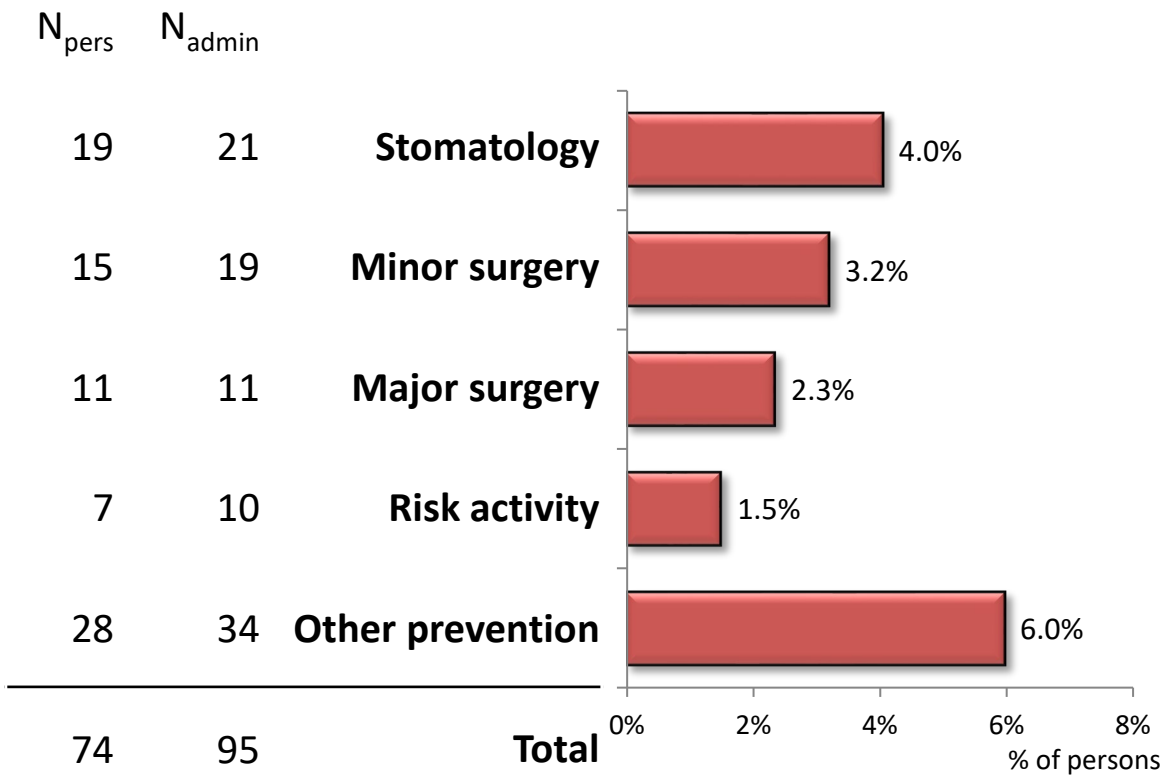
24 (9.7%) children were given factor to prevent bleeding during/before risk situation.
27 preventive administrations were recorded in total.



Preventive administration in 2022

Adults
Haem A
N=406

74 (18.2%) persons were given factor to prevent bleeding during/before risk situation.
95 preventive administrations were recorded in total.



ABR according to treatment regimen Haemophilia A without inhibitor

Annual bleeding rate according to treatment regimen

Children Haem A N=232*

Frequency of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*124*	*0*	*14*	*7*	*6*	*81*
Mean	*0.2*		*1.6*	*2.4*	***0.2***	***1.2***
Median (min – max)	*0 (0 – 7)*		*1 (0 – 10)*	*1 (0 – 10)*	***0 (0 – 1)***	***0 (0 – 7)***
Total no of recorded bleeds	*25*		*23*	*17*	***1***	***95***
Children on permanent prophylaxis	***0 (0%)***		***7 (33.3%)***		***81 (93.1%)***	
% of factor (FVIII) consumed by children on permanent prophylaxis	-		***83.8%***		***95.2%***	

Location of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*124*	*0*	*14*	*7*	*6*	*81*
JOINT BLEEDS						
Mean	*0.1*		*0.6*	*1.1*	***0.2***	***0.6***
Median (range)	*0 (0 – 1)*		*0 (0 – 3)*	*0 (0 – 8)*	***0 (0 – 1)***	***0 (0 – 5)***
Total no of recorded bleeds	*9*		*9*	*8*	***1***	***45***
OTHER BLEEDS						
Mean	*0.1*		*1.0*	*1.6*	***0.0***	***0.7***
Median (range)	*0 (0 – 6)*		*0 (0 – 9)*	*2 (0 – 3)*	***0 (0 – 0)***	***0 (0 – 4)***
Total no of recorded bleeds	*17*		*14*	*11*	***0***	***54***

Treatment regimen:
<u>OD</u> = on demand and/or temporary prophylaxis
<u>prophy</u> = permanent prophylaxis

** without inhibitor; missing location of their bleeds in 1 child (one of them is inhibitor)*

Annual bleeding rate according to treatment regimen

Adults
Haem A
N=392*

Frequency of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*217*	*0*	*30*	*4*	*32*	*109*
Mean	*0.1*	*0.0*	*0.7*	*0.8*	***5.6***	***1.1***
Median (min – max)	*0 (0 – 3)*	*(–)*	*0 (0 – 6)*	*0.5 (0 – 2)*	***0.5 (0 – 35)***	***0 (0 – 10)***
Total no of recorded bleeds	*17*	*0*	*21*	*3*	***178***	***115***
Adults on permanent prophylaxis	***0 (0%)***		***4 (11.4%)***		***109 (76.2%)***	
% of factor (FVIII) consumed by children on permanent prophylaxis	-		***75.0%***		***96.3%***	

Location of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*217*	*0*	*30*	*4*	*31*	*109*
JOINT BLEEDS						
Mean	*0.0*	*0*	*0.3*	*0.8*	***4.8***	***0.7***
Median (range)	*0 (0 – 3)*	*(–)*	*0 (0 – 5)*	*0.5 (0 – 2)*	***0 (0 – 26)***	***0 (0 – 9)***
Total no of recorded bleeds	*8*	*0*	*9*	*3*	***148***	***78***
OTHER BLEEDS						
Mean	*0.0*	*0*	*0.4*	*0.0*	***1.0***	***0.3***
Median (range)	*0 (0 – 1)*	*(–)*	*0 (0 – 6)*	*0 (0 – 0)*	***0 (0 – 9)***	***0 (0 – 5)***
Total no of recorded bleeds	*9*	*0*	*13*	*0*	***30***	***37***

Treatment regimen:
<u>OD</u> = on demand and/or temporary prophylaxis
<u>prophy</u> = permanent prophylaxis

** without inhibitor; missing severity in 1 adult; missing frequency of bleeding in 3 adults; missing location of bleeds in 1 adult*

ABR according to treatment regimen and age

Adults Haem A N=392*

** without inhibitor; missing severity in 1 adult; missing frequency of bleeding in 3 adults; missing location of bleeds in 1 adult*

Frequency of bleeding	Mild*		Moderate*		Severe*		
Treatment regimen	OD	Prophy	OD	Prophy	OD	Prophy	
N valid	161	0	19	2	26	76	Adults (haem A) born before 1990 N=284
Mean	0.0	0.0	0.7	1.0	**6.7**	**1.1**	
Median (min – max)	0 (0 – 1)	(–)	0 (0 – 6)	1 (0 – 2)	**3 (0 – 35)**	**0 (0 – 10)**	
Total no of recorded bleeds	6	0	14	2	**175**	**83**	
Adults on permanent prophylaxis	**0 (0%)**		**2 (9.5%)**		**76 (73.1%)**		
% of factor (FVIII) consumed by adults on permanent prophylaxis	**0.0%**		**63.3%**		**97.0%**		

Frequency of bleeding	Mild*		Moderate*		Severe*		
Treatment regimen	OD	Prophy	OD	Prophy	OD	Prophy	
N valid	56	0	11	2	6	33	Adults (haem A) born in 1990 or later N=108
Mean	0.2	0.0	0.6	0.5	**0.5**	**1.0**	
Median (min – max)	0 (0 – 3)	(–)	0 (0 – 6)	0.5 (0 – 1)	**0 (0 – 2)**	**0 (0 – 8)**	
Total no of recorded bleeds	11	0	7	1	**3**	**32**	
Adults on permanent prophylaxis	**0 (0%)**		**2 (14.3%)**		**33 (84.6%)**		
% of factor (FVIII) consumed by adults on permanent prophylaxis	**0.0%**		**91.4%**		**94.8%**		

Joint and other bleeds according to treatment regimen and age

Adults Haem A N=392*

** without inhibitor; missing severity in 1 adult; missing frequency of bleeding in 3 adults; missing location of bleeds in 1 adult*

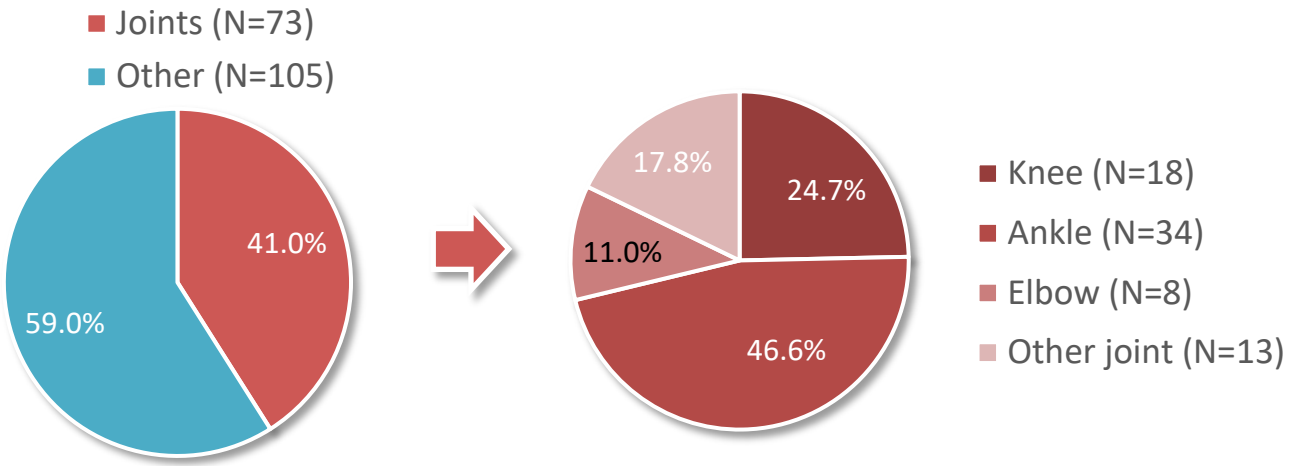
Frequency of bleeding	Mild*		Moderate*		Severe*		
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*	
N valid	*161*	*0*	*19*	*2*	*25*	*76*	
JOINT BLEEDS							
Mean	*0.0*	0	*0.5*	*1.0*	***5.9***	***0.8***	**Adults (haem A) born before 1990 N=281**
Median (range)	*0 (0 – 1)*	(–)	*0 (0 – 5)*	*1 (0 – 2)*	***2 (0 – 26)***	***0 (0 – 9)***	
Total no of recorded bleeds	*2*	0	*9*	*2*	***148***	***57***	
OTHER BLEEDS							
Mean	*0.0*	0	*0.3*	*0.0*	***1.1***	***0.3***	
Median (range)	*0 (0 – 1)*	(–)	*0 (0 – 2)*	*0 (0 – 0)*	***0 (0 – 9)***	***0 (0 – 5)***	
Total no of recorded bleeds	*4*	0	*6*	*0*	***27***	***26***	

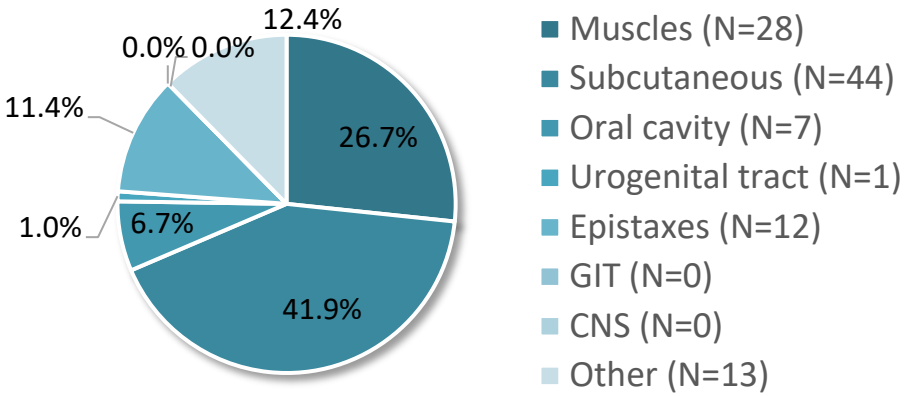
Frequency of bleeding	Mild*		Moderate*		Severe*		
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*	
N valid	*56*	*0*	*11*	*2*	*6*	*33*	
JOINT BLEEDS							
Mean	*0.1*	0	*0.0*	*0.5*	***0.0***	***0.6***	**Adults (haem A) born in 1990 or later N=108**
Median (range)	*0 (0 – 3)*	(–)	*0 (0 – 0)*	*0.5 (0 – 1)*	***0 (0 – 0)***	***0 (0 – 6)***	
Total no of recorded bleeds	*6*	0	*0*	*1*	***0***	***21***	
OTHER BLEEDS							
Mean	*0.1*	0	*0.6*	*0.0*	***0.5***	***0.3***	
Median (range)	*0 (0 – 1)*	(–)	*0 (0 – 6)*	*0 (0 – 0)*	***0 (0 – 2)***	***0 (0 – 5)***	
Total no of recorded bleeds	*5*	0	*7*	*0*	***3***	***11***	

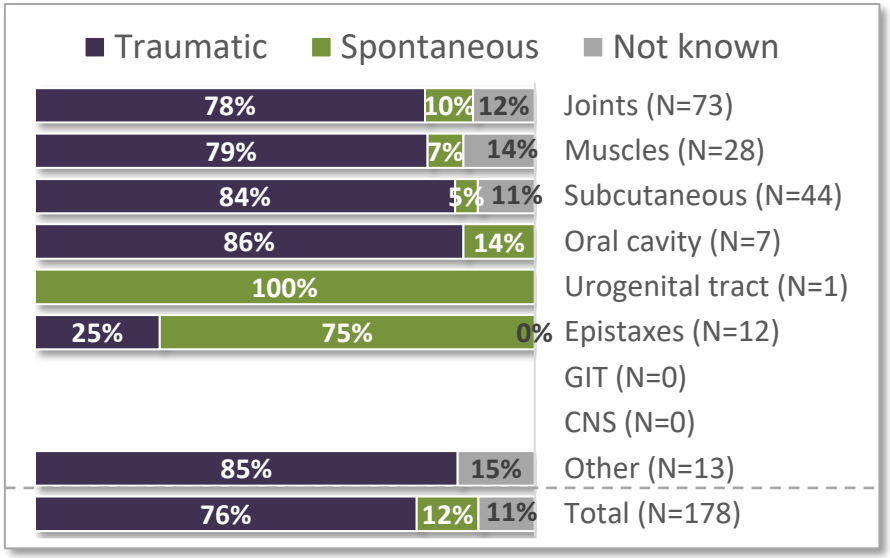
Location and etiology of bleeds

Children Haem A N=178*

** number of bleeds*







Detailed treatment of bleeds

Children Haem A N=178*

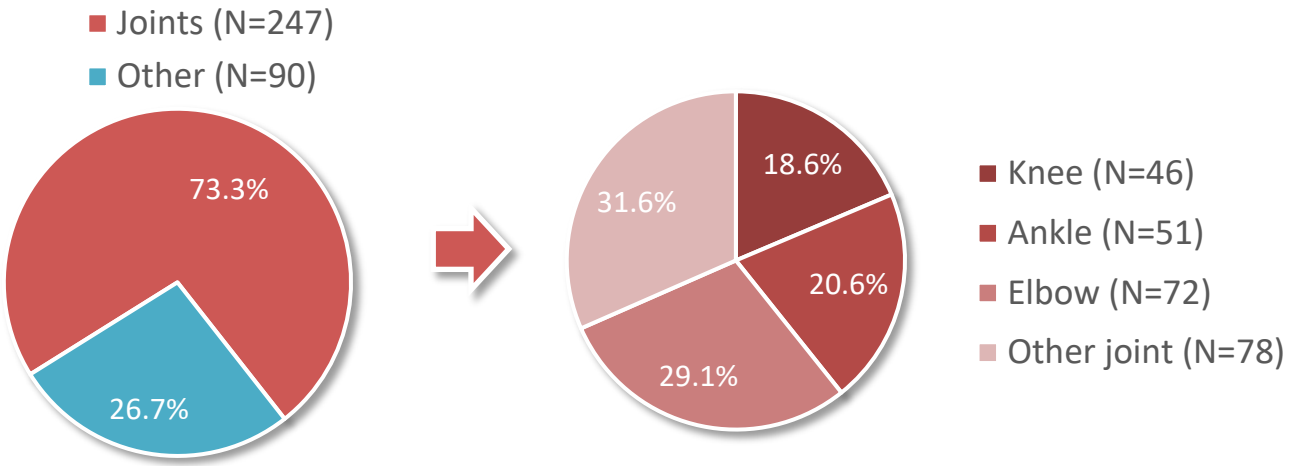
** number of bleeds*

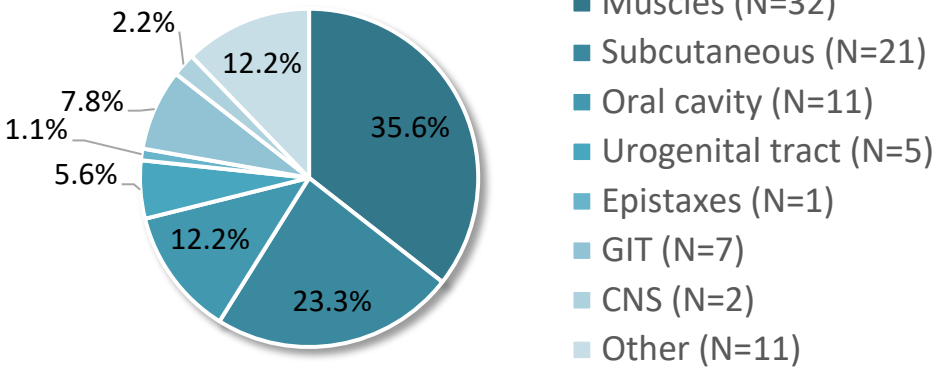
	Joints	Muscles	Subcuta-neous	Oral cavity	Urogenital tract	Epistaxes	GIT	CNS	Other	Total
No. of bleeds	*73*	*28*	*44*	*7*	*1*	*12*	*0*	*0*	*13*	***178***
FVIII consumption per bleed (IU), valid N	*62*	*25*	*36*	*4*	*1*	*4*			*12*	***144***
geometric mean	2 376.5	2 659.6	976.4	707.1	1 000.0	1 029.9			1 601.0	**1 762.9**
median	**2 000.0**	**2 000.0**	**1 000.0**	**1 125.0**	**1 000.0**	**1 000.0**			**1 875.0**	**1 500.0**
min – max	250–28000	250–22500	250–5000	250–2000	1000–1000	750–1500			250–12000	**250–28000**
sum	233 000	123 250	47 501	4 500	1000	4 250			29 500	**443 001**
No. of doses per bleed										
geometric mean	1.6	2.0	1.3	1.2	1.0	1.0			1.5	**1.6**
median	**1**	**2**	**1**	**1**	**1**	**0**			**1**	**1**
min – max	0–11	0–14	0–3	0–2	1–1	0–1			0–12	**0–14**
Duration of therapy per bleed, days										
geometric mean	1.9	2.3	1.4	1.6	1.0	1.5			1.7	**1.8**
median	**1**	**3**	**1**	**1**	**1**	**1**			**1**	**1**
min – max	1–20	1–18	1–4	1–10	1–1	1–7			1–20	**1–20**
N (%) with hospitalization	*6 (8.2%)*	*2 (7.1%)*	*1 (2.3%)*	*1 (14.3%)*	*0 (0%)*	*1 (8.3%)*			*1 (7.7%)*	***12 (6.7%)***
N (%) with rebleeding	*10 (13.7%)*	*1 (3.6%)*	*7 (15.9%)*	*0 (0%)*	*0 (0%)*	*7 (58.3%)*			*1 (7.7%)*	***26 (14.6%)***

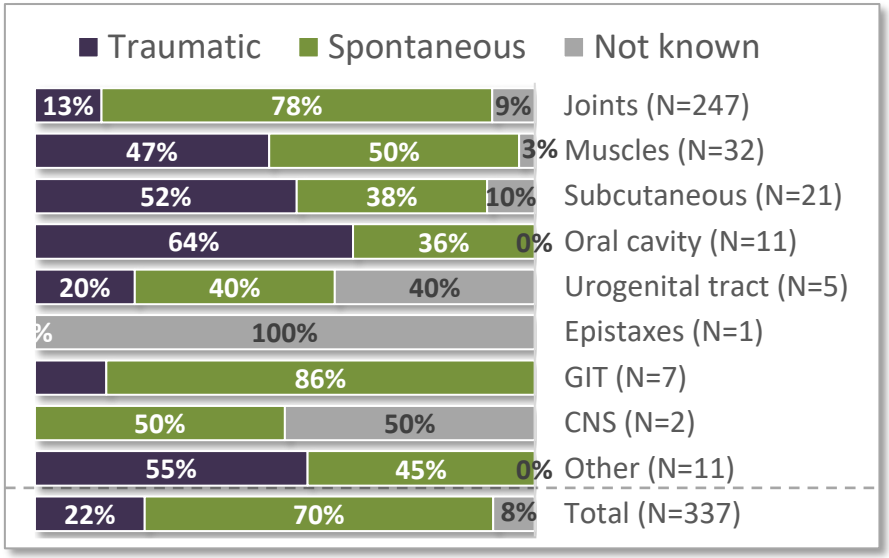
Location and etiology of bleeds

Adults
Haem A
N=337*

** number of bleeds*







Detailed treatment of bleeds

Adults
Haem A
N=337*

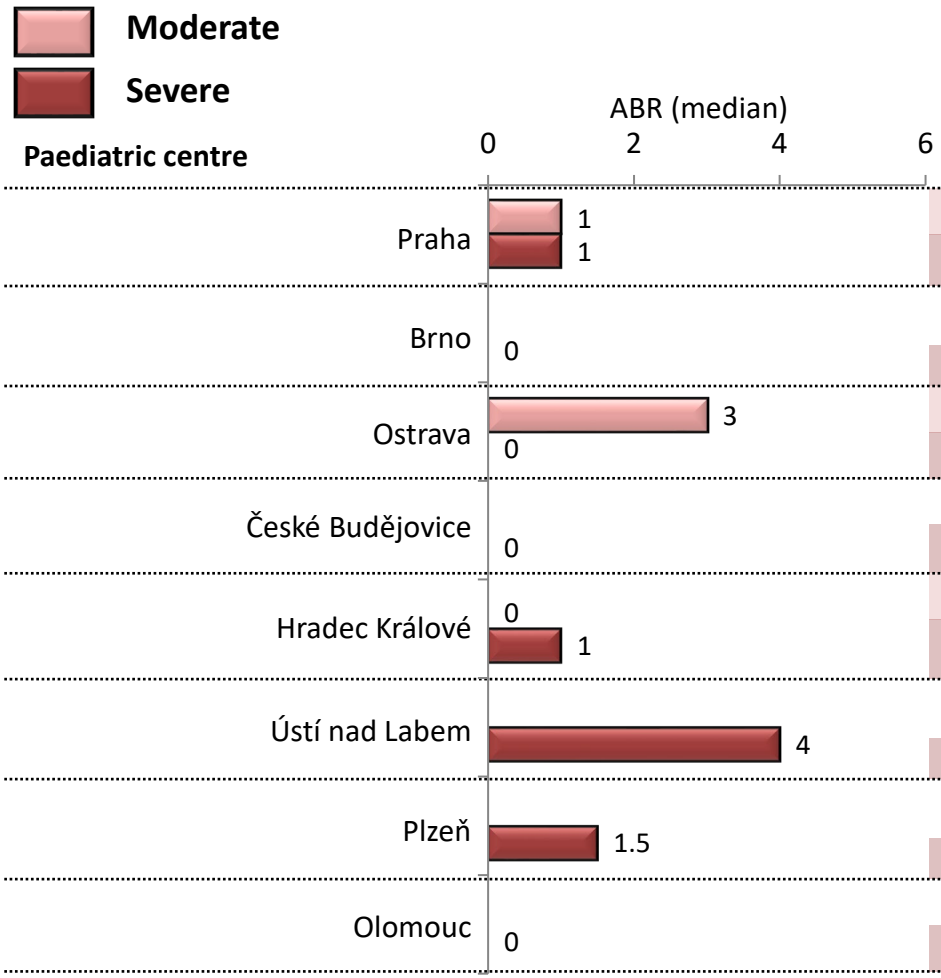
** number of bleeds*

	Joints	Muscles	Subcuta-neous	Oral cavity	Urogenital tract	Epistaxes	GIT	CNS	Other	Total
No. of bleeds	*247*	*32*	*21*	*11*	*5*	*1*	*7*	*2*	*11*	***337***
FVIII consumption per bleed (IU), valid N	*244*	*32*	*21*	*11*	*4*	*1*	*7*	*2*	*10*	***332***
geometric mean	2035.4	2400.1	3938.8	2832.7	6636.7	2000.0	28571.5	93179.9	4115.9	**2443.4**
median	**2000.0**	**2000.0**	**4000.0**	**2000.0**	**6000.0**	**2000.0**	**17000.0**	**104250.0**	**4000.0**	**2000.0**
min – max	500–20000	500–166000	500–53500	1000–17000	2000–48500	2000–2000	500–1776000	57500–151000	500–19000	**500–1 776 000**
sum	723 500	324 500	143 000	50 500	62 500	2 000	2 098 000	208 500	69 500	**3 682 000**
No. of doses per bleed										
geometric mean	1.5	1.9	1.6	1.7	2.7	1.0	12.1	61.3	1.8	**1.7**
median	**1**	**1**	**1**	**1**	**2**	**1**	**9**	**62**	**1**	**1**
min – max	0–2000	1–80	1–24	1–7	1–24	1–1	1–75	53–71	0–8	**0–2000**
Duration of therapy per bleed, days										
geometric mean	1.5	1.8	1.6	1.7	2.1	1.0	8.1	30.0	1.8	**1.6**
median	**1**	**1**	**1**	**1**	**2**	**1**	**8**	**31**	**1**	**1**
min – max	1–10	1–51	1–17	1–9	1–10	1–1	1–60	25–36	1–8	**1–60**
N (%) with hospitalization	*2 (0.8%)*	*4 (12.5%)*	*2 (9.5%)*	*0 (0%)*	*1 (20%)*	*0 (0%)*	*6 (85.7%)*	*2 (100%)*	*0 (0%)*	***17 (5%)***
N (%) with rebleeding	*3 (1.2%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	***3 (0.9%)***

ABR according to centres Haemophilia A (PWHA)

Annual bleeding rate on permanent prophylaxis

HaemA on prophy
Paed. centres
N=96

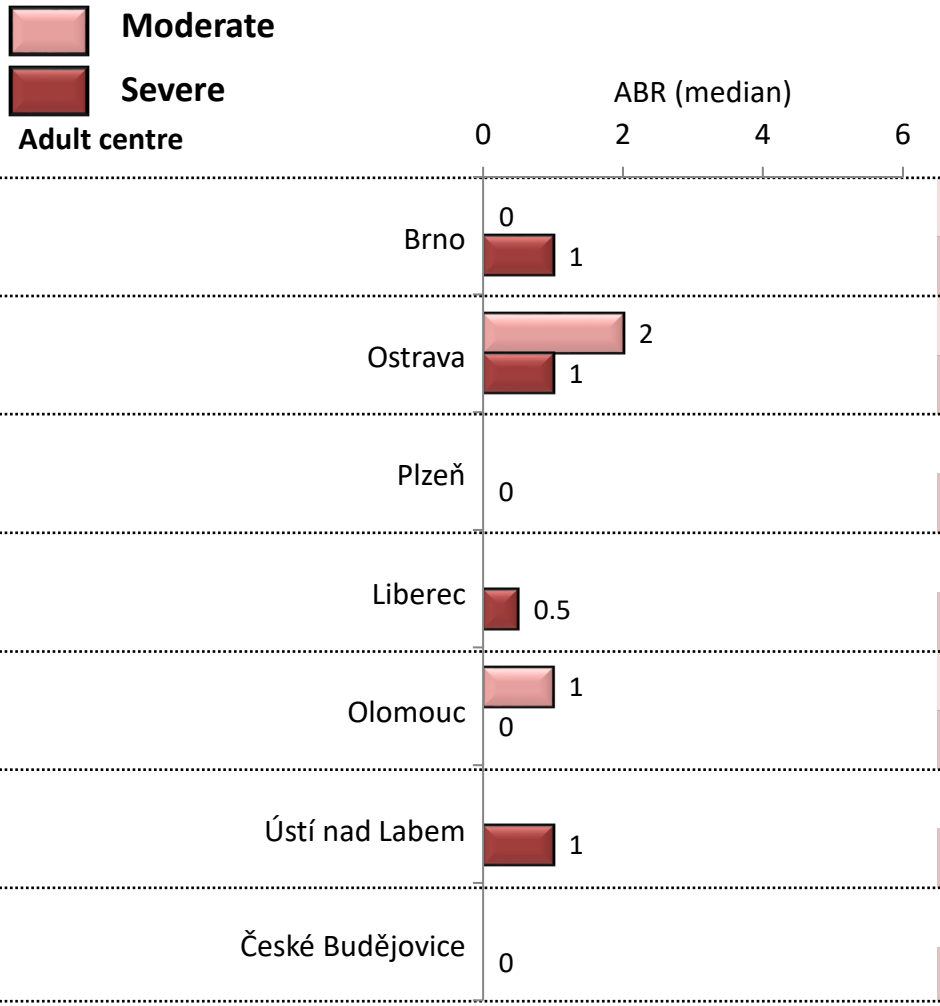


Frequency of bleeding in PWHA without inhibitor on permanent prophylaxis

Paediatric centre	N	Mean	Median	Min	Max	Severity
Praha	5	2.8	1.0	0	10	Moderate
	35	1.7	1.0	0	7	Severe
Brno	0					
	21	0.8	0.0	0	6	Severe
Ostrava	1	3.0	3.0	3	3	Moderate
	10	0.4	0.0	0	1	Severe
České Budějovice	0					
	11	0.3	0.0	0	1	Severe
Hradec Králové	2	0.0	0.0	0	0	Moderate
	3	0.7	1.0	0	1	Severe
Ústí nad Labem	0					
	3	3.0	4.0	0	5	Severe
Plzeň	0					
	2	1.5	1.5	1	2	Severe
Olomouc	0					
	3	0.0	0.0	0	0	Severe

Annual bleeding rate on permanent prophylaxis

HaemA on prophy
Adult centres
N=105



Frequency of bleeding in PWHA without inhibitor on permanent prophylaxis

Adult centre	N	Mean	Median	Min	Max	Severity
Brno	1	0.0	0.0	0	0	Moderate
	33	1.1	1.0	0	8	Severe
Ostrava	1	2.0	2.0	2	2	Moderate
	24	1.8	1.0	0	7	Severe
Plzeň	0					
	16	0.3	0.0	0	4	Severe
Liberec	0					
	6	1.2	0.5	0	3	Severe
Olomouc	1	1.0	1.0	1	1	Moderate
	10	1.3	0.0	0	10	Severe
Ústí nad Labem	0					
	7	0.9	1.0	0	2	Severe
České Budějovice	0					
	6	0.0	0.0	0	0	Severe

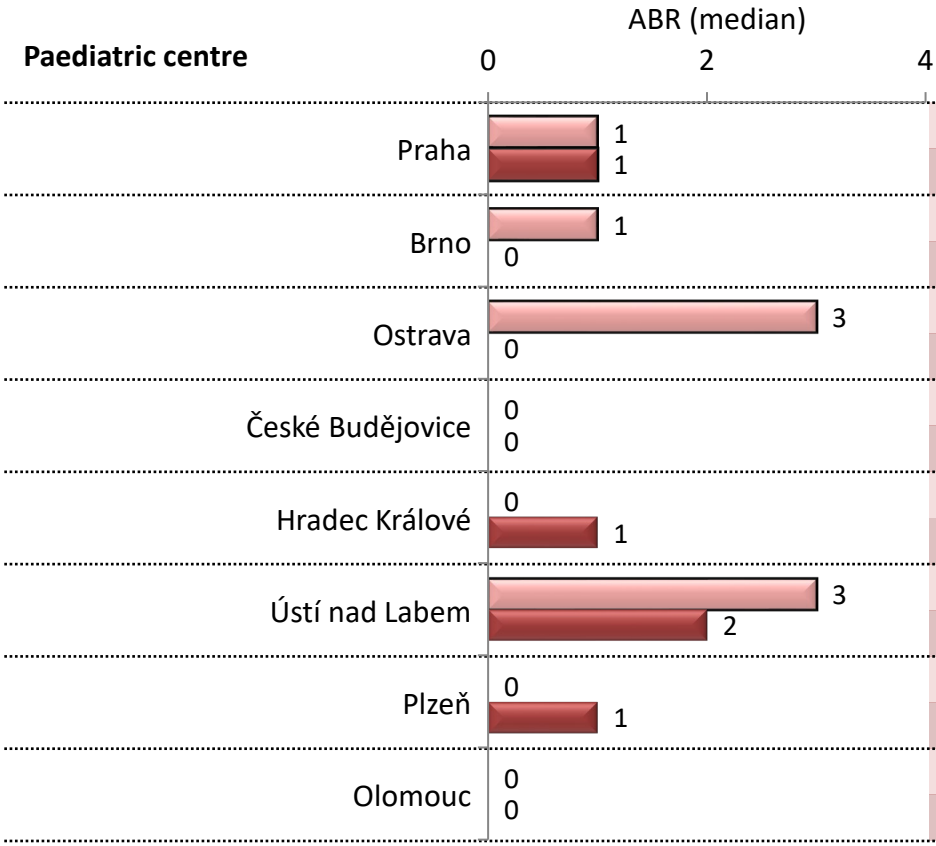
Annual bleeding rate regardless prophylaxis

HaemA
Paed. centres
N=118

Moderate

Severe

Frequency of bleeding in PWHA without inhibitor regardless of prophylaxis



Paediatric centre	N	Mean	Median	Min	Max	% on permanent prophylaxis
Praha	8	2.1	1.0	0	10	62.5%
	37	1.7	1.0	0	7	94.6%
Brno	5	0.6	1.0	0	1	0.0%
	22	0.7	0.0	0	6	95.5%
Ostrava	1	3.0	3.0	3	3	100.0%
	10	0.4	0.0	0	1	100.0%
České Budějovice	3	1.3	0.0	0	4	0.0%
	12	0.3	0.0	0	1	91.7%
Hradec Králové	2	0.0	0.0	0	0	100.0%
	3	0.7	1.0	0	1	100.0%
Ústí nad Labem	1	3.0	3.0	3	3	0.0%
	4	2.3	2.0	0	5	75.0%
Plzeň	1	0.0	0.0	0	0	0.0%
	3	1.0	1.0	0	2	66.7%
Olomouc	3	3.3	0.0	0	10	0.0%
	3	0.0	0.0	0	0	100.0%

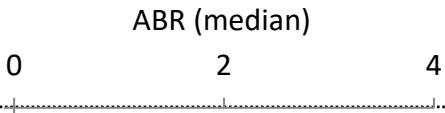
Annual bleeding rate regardless prophylaxis

HaemA
Adult centres
N=168*

** missing ABR in 3 adults*

Moderate
Severe

Frequency of bleeding in PWHA without inhibitor regardless of prophylaxis



Adult centre	N	Mean	Median	Min	Max	% on permanent prophylaxis
Brno	14	0.4	0.0	0	1	7.1%
	41	1.1	0.0	0	8	80.5%
Ostrava	6	1.5	0.5	0	6	16.7%
	28	2.4	1.0	0	13	85.7%
Plzeň	2	0.0	0.0	0	0	0.0%
	21	0.4	0.0	0	4	80.0%
Liberec	3	0.7	1.0	0	1	0.0%
	11	4.6	1.0	0	24	54.5%
Olomouc	1	1.0	1.0	1	1	50.0%
	18	6.1	0.5	0	35	50.0%
Ústí nad Labem	3	0.0	0.0	0	0	0.0%
	7	0.9	1.0	0	2	100.0%
České Budějovice	3	2.0	0.0	0	6	0.0%
	10	0.3	0.0	0	3	66.7%

Prophylactic regimens and treatment outcomes

HaemA
Paed. centres
N=118

Paediatric centre	Severity	Total N	PERMANENT PROPHYLAXIS										ON-DEMAND / TEMPORARY PROPHY		
			% of patients	N	Dosing of prophylaxis SHL (IU/kg per week)		Dosing of prophylaxis EHL (IU/kg per week)		Dosing of EMI prophylaxis (mg/kg per week)		ABR		N	ABR	
					Mean	Median	Mean	Median	Mean	Median	Mean	Median		Mean	Median
Praha	Moderate	8	**62.5%**	5	67.7	**64.5**	49.3	**49.3**	1.6	**1.6**	2.8	**1.0**	3	1.0	1.0
	Severe	37	**94.6%**	35	86.9	**100.0**	66.4	**65.3**	1.5	**1.5**	1.7	**1.0**	2	0.5	0.5
Brno	Moderate	5	**0.0%**	0									5	0.6	1.0
	Severe	22	**95.5%**	21			79.2	**80.7**	1.2	**1.1**	0.8	**0.0**	1	0.0	0.0
Ostrava	Moderate	1	**100.0%**	1	73.8	**73.8**					3.0	**3.0**	0		
	Severe	10	**100.0%**	10	44.1	**44.1**	74.6	62.4	1.5	1.5	0.4	**0.0**	0		
Č. Budějovice	Moderate	3	**0.0%**	0									3	1.3	0.0
	Severe	12	**91.7%**	11	82.4	**82.4**	72.1	**72.3**			0.3	**0.0**	1	0.0	0.0
Hradec Králové	Moderate	2	**100.0%**	2			42.9	**42.9**			0.0	**0.0**	0		
	Severe	3	**100.0%**	3			80.4	**64.7**			0.7	**1.0**	0		
Ústí nad Labem	Moderate	1	**0.0%**	0									1	3.0	3.0
	Severe	4	**75.0%**	3	75.0	**75.0**	67.7	**67.3**	1.6	**1.6**	3.0	**4.0**	1	0.0	0.0
Plzeň	Moderate	1	**0.0%**	0									1	0.0	0.0
	Severe	3	**66.7%**	2			101.7	**101.7**	1.5	**1.5**	1.5	**1.5**	1	0.0	0.0
Olomouc	Moderate	3	**0.0%**	0									3	3.3	0.0
	Severe	3	**100.0%**	3			21.7	**21.7**	1.5	**1.5**	0.0	**0.0**	0		

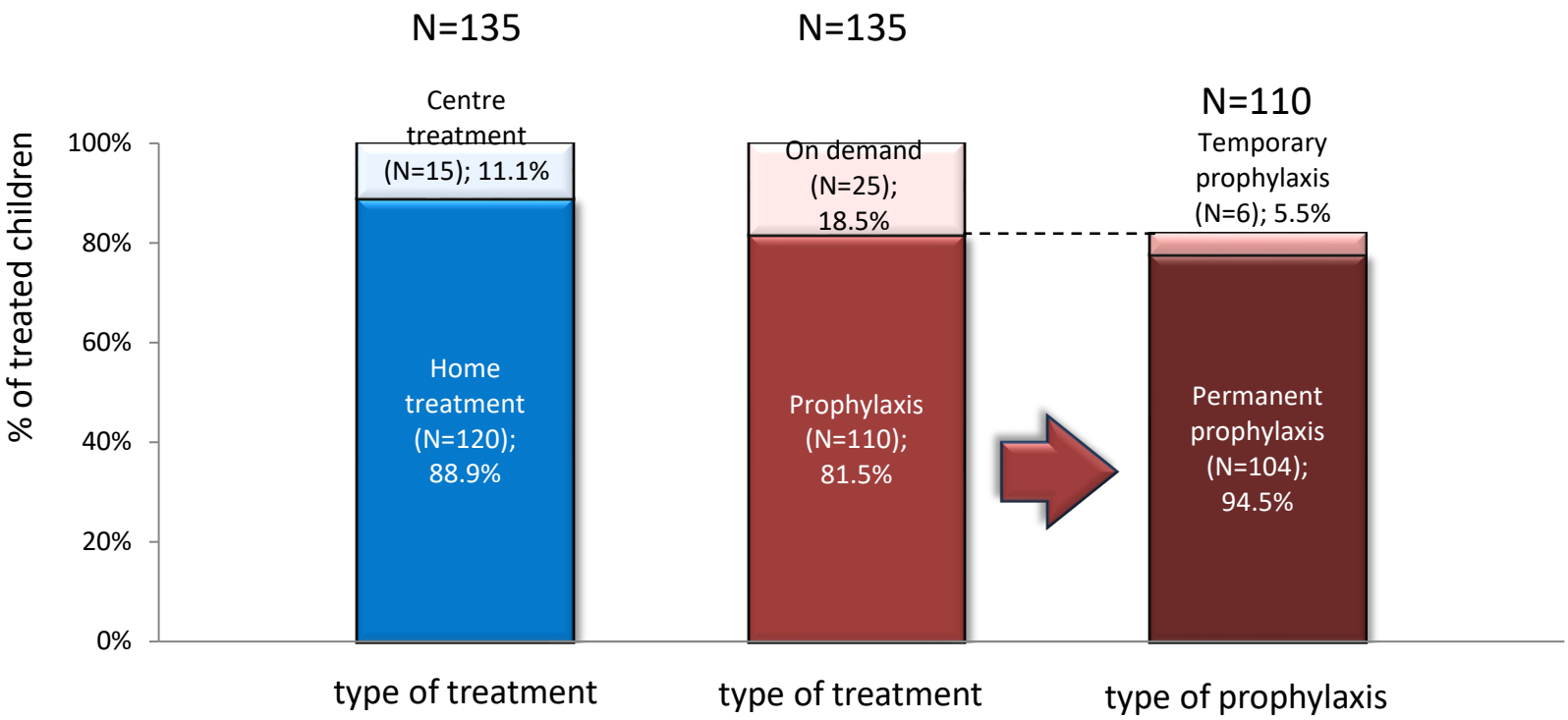
Prophylactic regimens and treatment outcomes

HaemA
Adult centres
N=171

Adult centre	Severity	Total N	PERMANENT PROPHYLAXIS											ON-DEMAND / TEMPORARY PROPHY			
			% of patients	N	Dosing of prophylaxis SHL (IU/kg per week)		Dosing of prophylaxis EHL (IU/kg per week)		Dosing of EMI prophylaxis (mg/kg per week)		ABR		Age	N	ABR		Age
					Mean	Median	Mean	Median	Mean	Median	Mean	Median	Median		Mean	Median	Median
Brno	**Moderate**	14	**7.1%**	1			50.0	**50.0**			0.0	**0.0**	33	13	0.5	0.0	48
	Severe	41	**80.5%**	33	75.2	**80.0**	71.1	**71.4**	1.5	**1.5**	1.1	**1.0**	38	8	1.0	0.0	47
Ostrava	**Moderate**	6	**16.7%**	1			72.4	**72.4**			2.0	**2.0**	70	5	1.4	0.0	33
	Severe	28	**85.7%**	24	61.8	**57.9**	73.6	**80.8**	1.5	**1.5**	1.8	**1.0**	43	4	5.5	4.5	60
Plzeň	**Moderate**	2	**0.0%**	0										2	0.0	0.0	41
	Severe	21	**80.0%**	16	47.4	**48.1**	55.2	**53.3**	2.9	**2.9**	0.3	**0.0**	47	4	1.0	0.0	57
Liberec	**Moderate**	3	**0.0%**	0										2	1.0	1.0	50
	Severe	11	**54.5%**	6	90.5	**90.5**	71.3	**72.0**			1.2	**0.5**	34	5	8.8	8.0	67
Olomouc	**Moderate**	2	**50.0%**	1	74.1	**74.1**					1.0	**1.0**	23	1	0.0	0.0	27
	Severe	20	**50.0%**	10	57.5	**59.2**	41.9	**45.0**			1.3	**0.0**	35	10	12.1	14.5	70
Ústí n. Labem	**Moderate**	3	**0.0%**	0										3	0.0	0.0	25
	Severe	7	**100.0%**	7			58.1	**58.9**			0.9	**1.0**	37	0			
Č. Budějovice	**Moderate**	3	**0.0%**	0										3	2.0	0.0	69
	Severe	10	**66.7%**	6			48.2	**55.2**			0.0	**0.0**	54	3	1.0	0.0	51

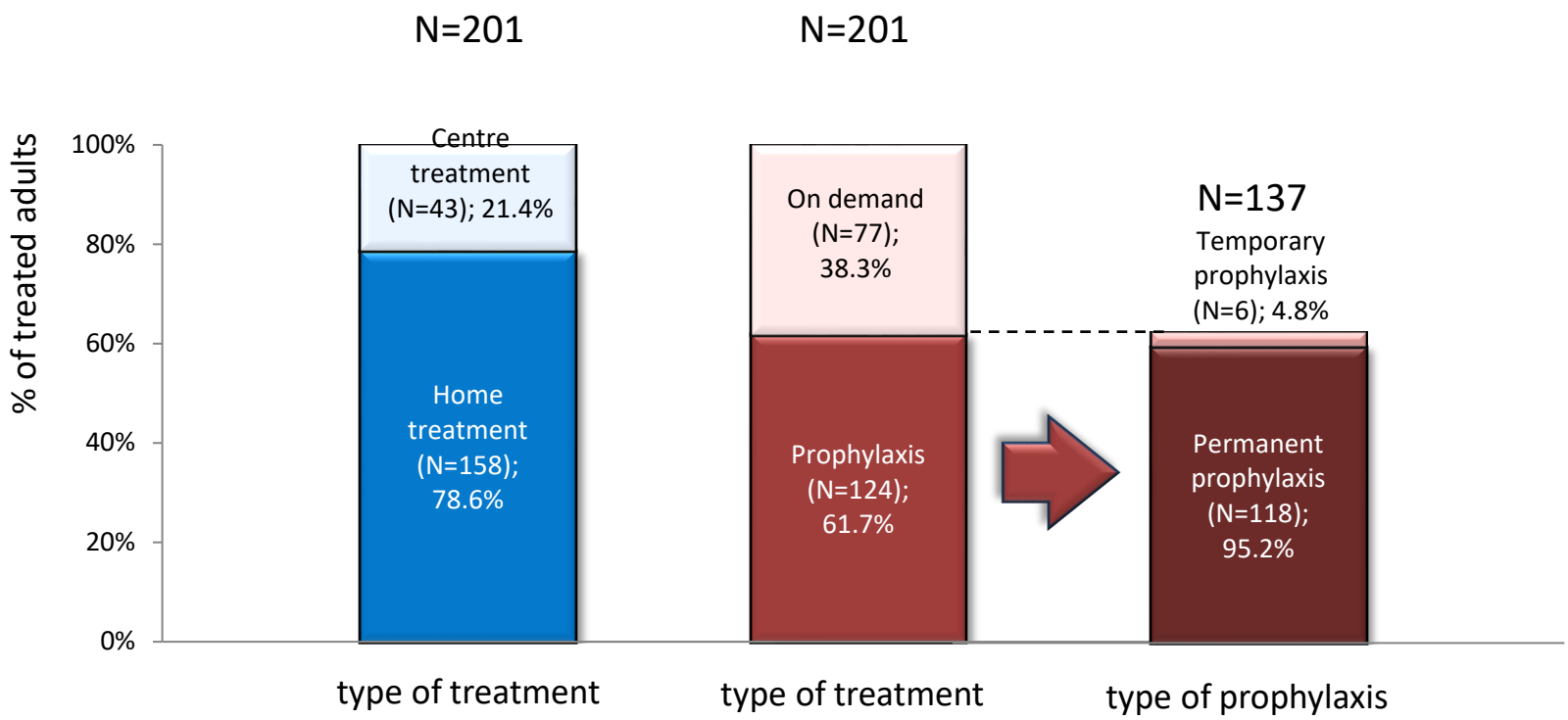
Children
Haem A
N=135

Type of treatment (subgroup of treated patients)



Adults
Haem A
N=201

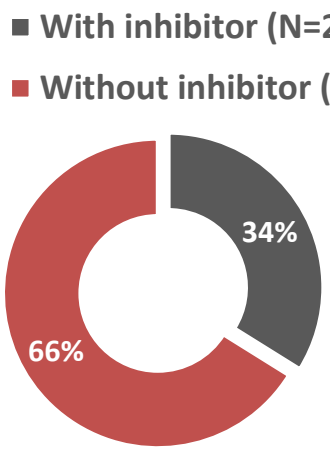
Type of treatment (subgroup of treated patients)

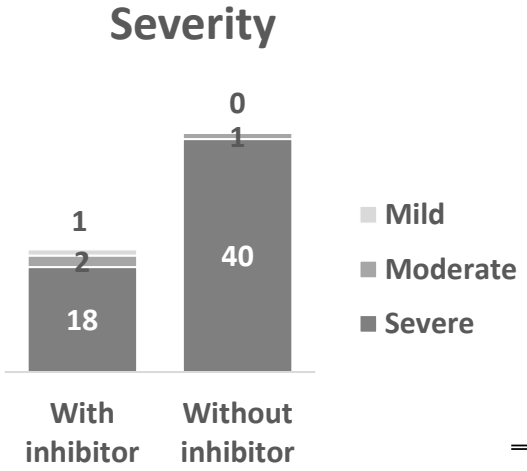


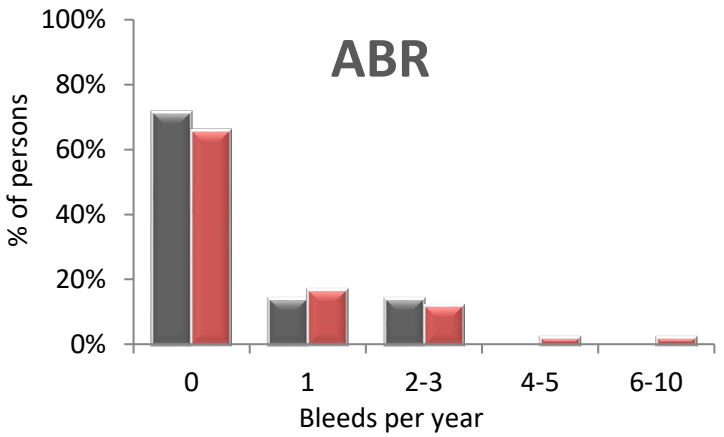
Emicizumab treatment in 2022

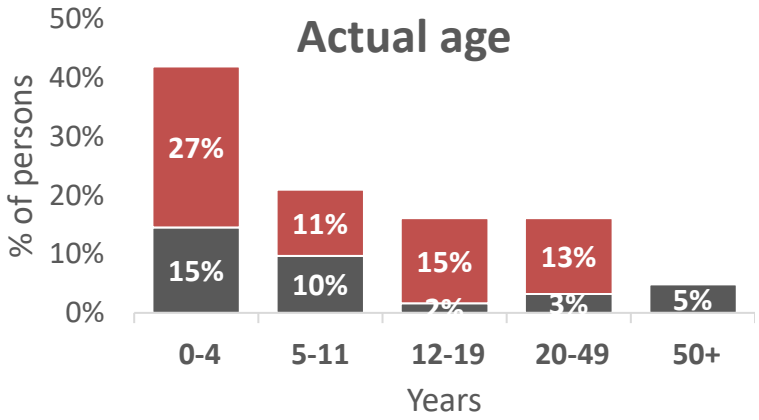
Haem A
N=62[1]

[1] *patients on emicizumab prophylaxis in a given year*









	Total	With inhibitor	Without inhibitor
N valid	*62*	*21*	*41*
ABR			
Mean	*0.61*	*2*	*0.3*
Median (min – max)	*0 (0 – 6)*	*0 (0 – 3)*	*0 (0 – 6)*
N (%) with no bleed	*42 (68%)*	*15 (71%)*	*27 (66%)*
EMI DOSE (mg/kg/week)			
Mean	*1.52*	*1.65*	*1.45*
Median (min – max)	*1.5 (0.9 – 3.4)*	*1.5 (1.2 – 3.4)*	*1.5 (0.9 – 2.9)*

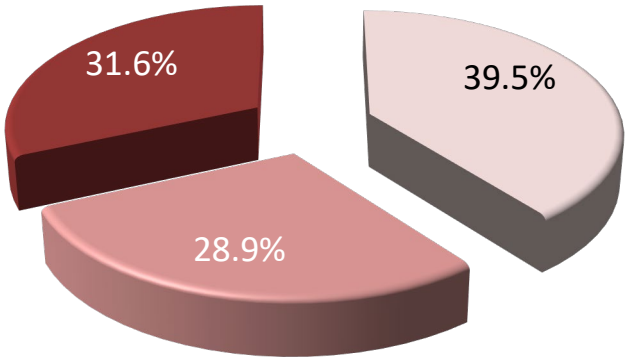
Demographic characteristics Haemophilia B

Severity of haemophilia B

Haem B
N=103

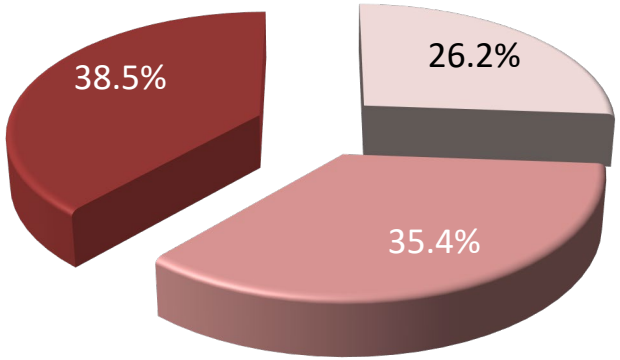
Children (N=38)

- Mild (N=15)
- Moderate (N=11)
- Severe (N=12)



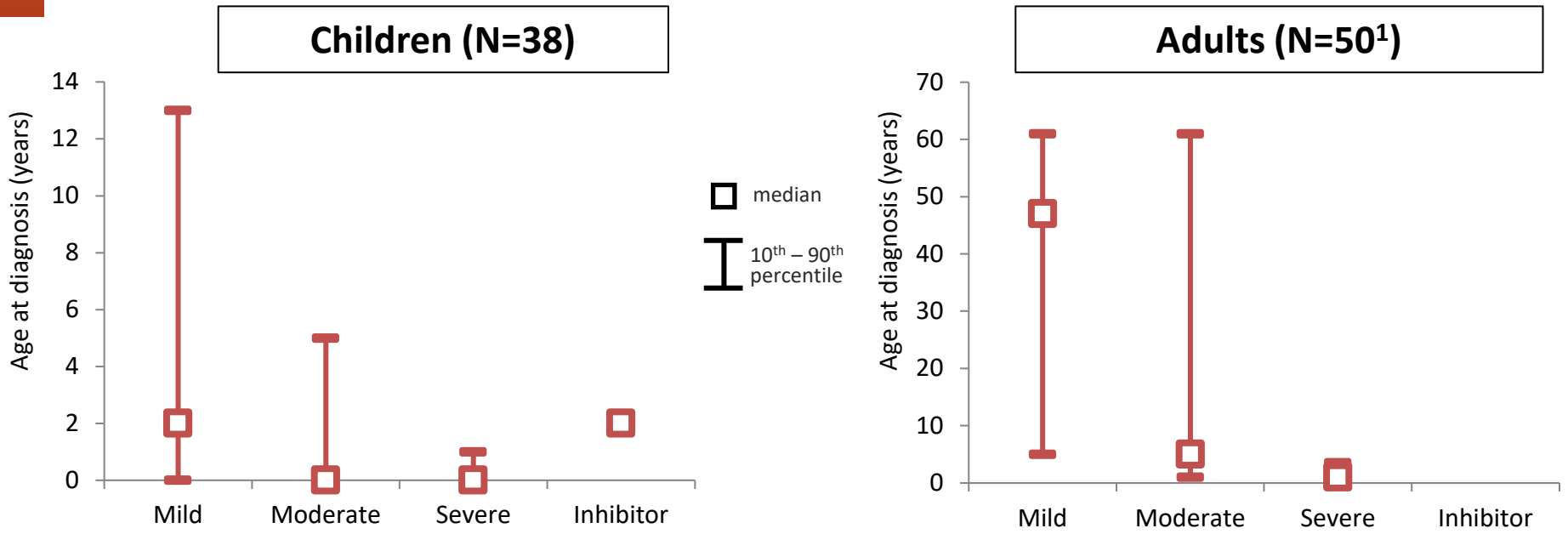
Adults (N=65)

- Mild (N=17)
- Moderate (N=23)
- Severe (N=25)



Age at diagnosis according to severity of haemophilia B

Haem B
N=103



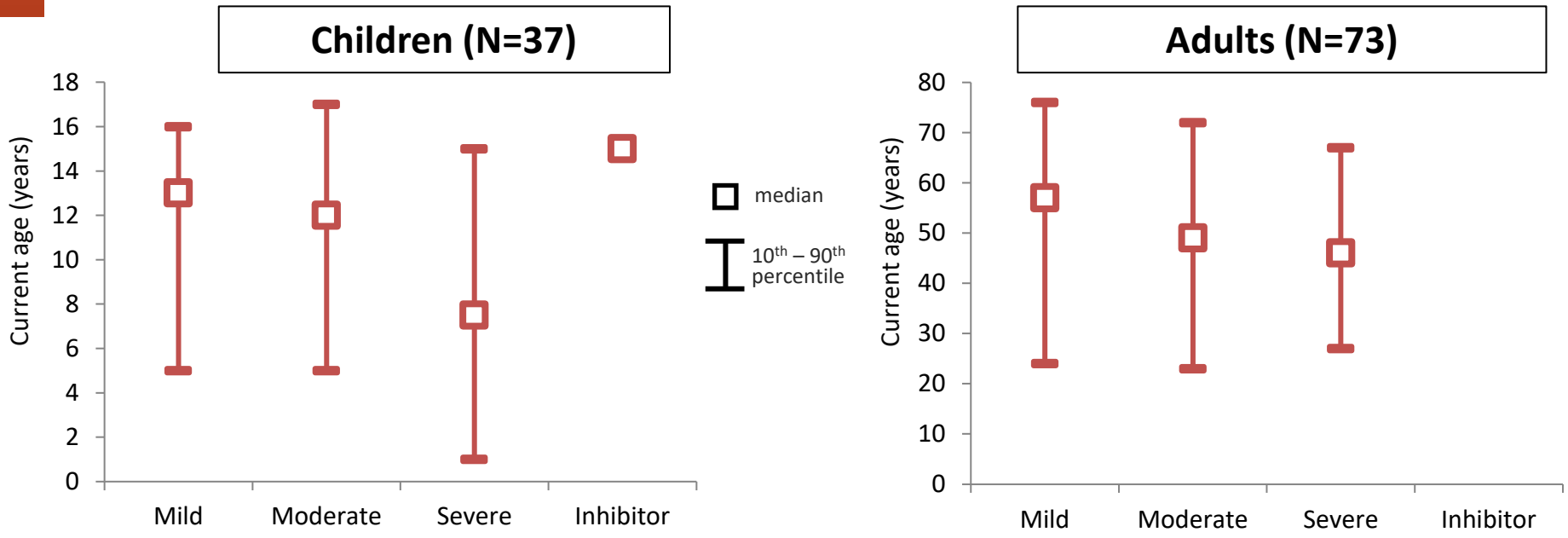
Mild*	Moderate*	Severe*	Inhibitor+	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor+
15	*11*	*12*	*1*	**N valid**	*11*	*19*	*20*	*0*
4.3	*1.9*	*0.5*	*2.0*	**Mean**	*33.7*	*12.3*	*1.4*	
2 (0 – 15)	*0 (0 – 8)*	*0 (0 – 2)*	*2 (2 – 2)*	**Median (min – max)**	*47 (2 – 67)*	*5 (0 – 69)*	*1 (0 – 4)*	

[1] *Missing information on year of diagnosis in 15 adults.*

** including persons with inhibitor*
+ in 2022

Haem B
N=103

Actual age according to severity of haemophilia B



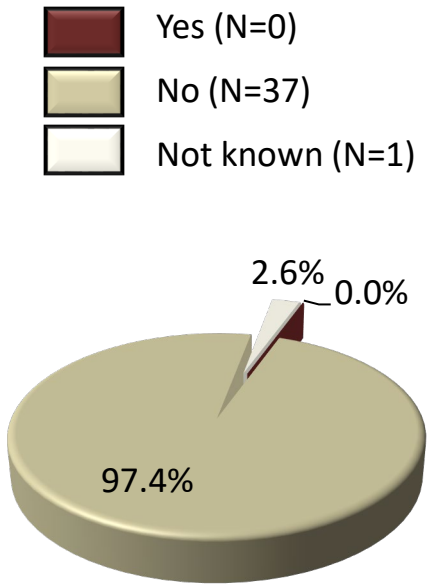
Mild*	Moderate*	Severe*	Inhibitor+	Current age++ (years)	Mild*	Moderate*	Severe*	Inhibitor+
15	*11*	*12*	*1*	**N valid**	*17*	*23*	*25*	*0*
11.4	*11.6*	*7.9*	*15.0*	**Mean**	*50.7*	*49.2*	*45.1*	
13 (4 – 18)	*12 (3 – 18)*	*7.5 (0 – 18)*	*15 (15 – 15)*	**Median (min – max)**	*57 (21 – 84)*	*49 (19 – 83)*	*46 (19 – 71)*	

** including persons with inhibitor*
+ in 2022
++ age reached in year 2022

Hepatitis (ever) experienced

Children
Haem B
N=38

Experienced hepatitis



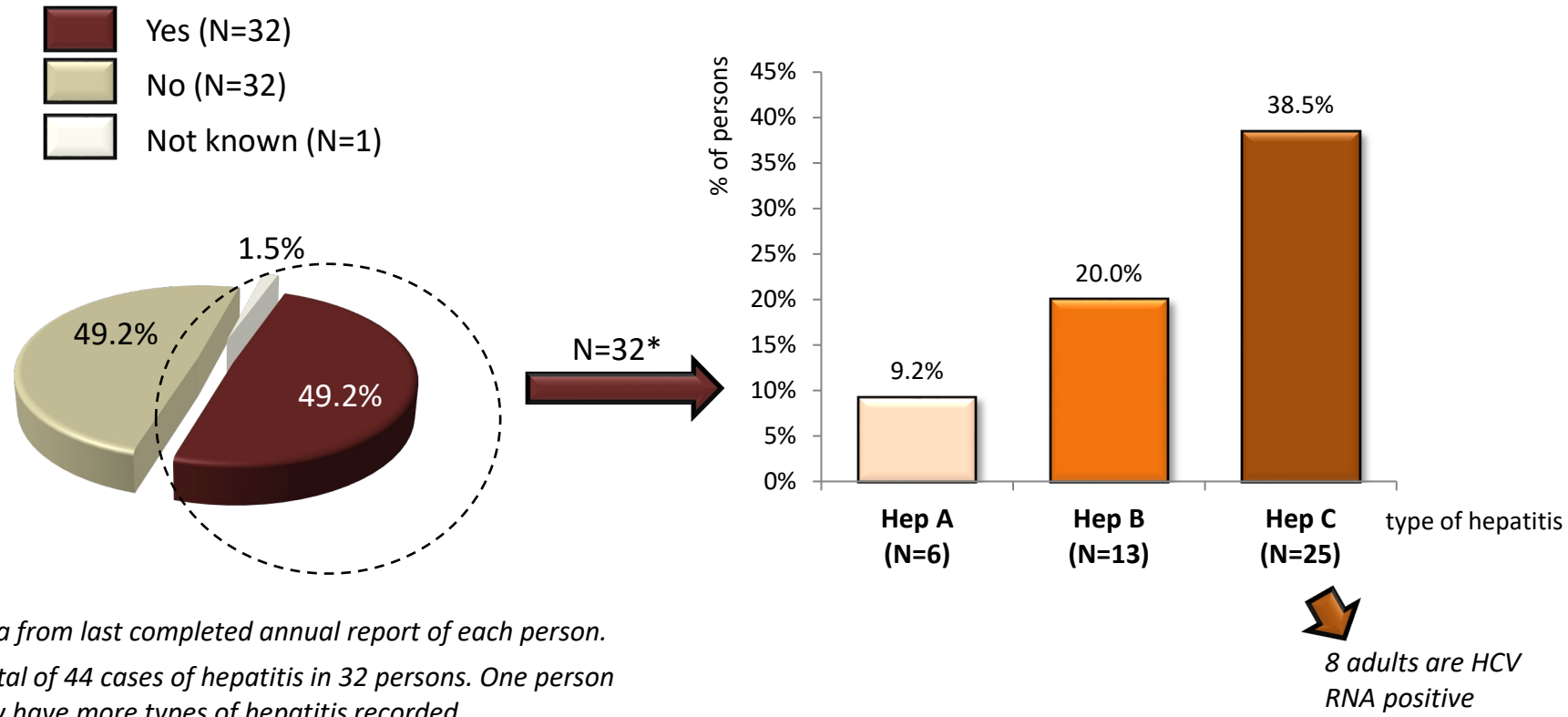
No child has hepatitis.

Data from last completed annual report of each person.

Hepatitis (ever) experienced

Adults
Haem B
N=65

Experienced hepatitis



Data from last completed annual report of each person.

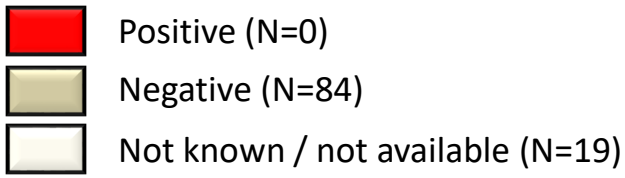
**Total of 44 cases of hepatitis in 32 persons. One person may have more types of hepatitis recorded.*

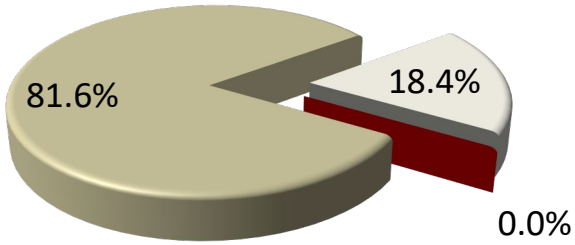
8 adults are HCV RNA positive

HIV

All
Haem B
N=103

HIV





No HIV-positive person.

Data from last completed annual report of each person.

Treatment outcomes and bleeding frequency Haemophilia B

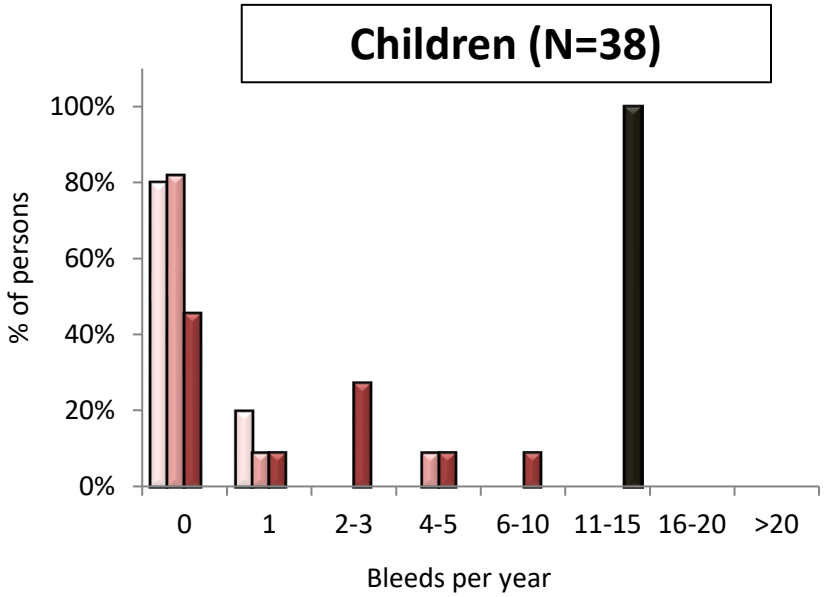
All
Haem B
N=103

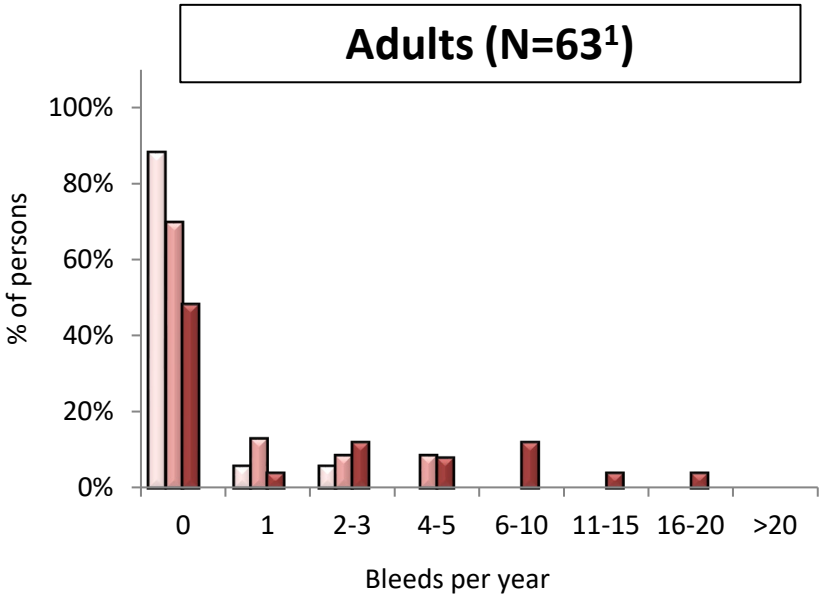
Data from year 2022 – sample size

	Valid persons			Persons with valid annual report			Persons examined			Persons treated	
	N	%		N	%		N	%		N	%
All	103	100%	→	100	97.1%	→	80	77.7%	→	58	56.3%
of them with inhibitor	1			1			1			1	
Children	38	100%	→	38	100.0%	→	32	84.2%	→	17	44.7%
of them with inhibitor	1			1			1			1	
Adults	65	100%	→	62	95.4%	→	48	73.8%	→	41	63.1%
of them with inhibitor	0			0			0			0	

Haem B
N=101[1]

Frequency of bleeding requiring treatment in 2022





Mild*	Moderate*	Severe*	Inhibitor	Frequency of bleeding	Mild*	Moderate*	Severe*	Inhibitor
15	*11*	*11*	*1*	**N valid**	*17*	*23*	*23*	*0*
0.2	*0.5*	*1.9*	*11.0*	**Mean**	*0.2*	*0.7*	*2.8*	*0.0*
0 (0 – 1)	*0 (0 – 4)*	*1 (0 – 9)*	*11 (11 – 11)*	**Median (min – max)**	*0 (0 – 2)*	*0 (0 – 4)*	*0 (0 – 18)*	*(–)*
12 (80%)	*9 (81.8%)*	*5 (45.5%)*	*0 (0%)*	**N (%) with no bleed**	*15 (88.2%)*	*16 (69.6%)*	*12 (48%)*	*0 (0%)*

** without inhibitor*

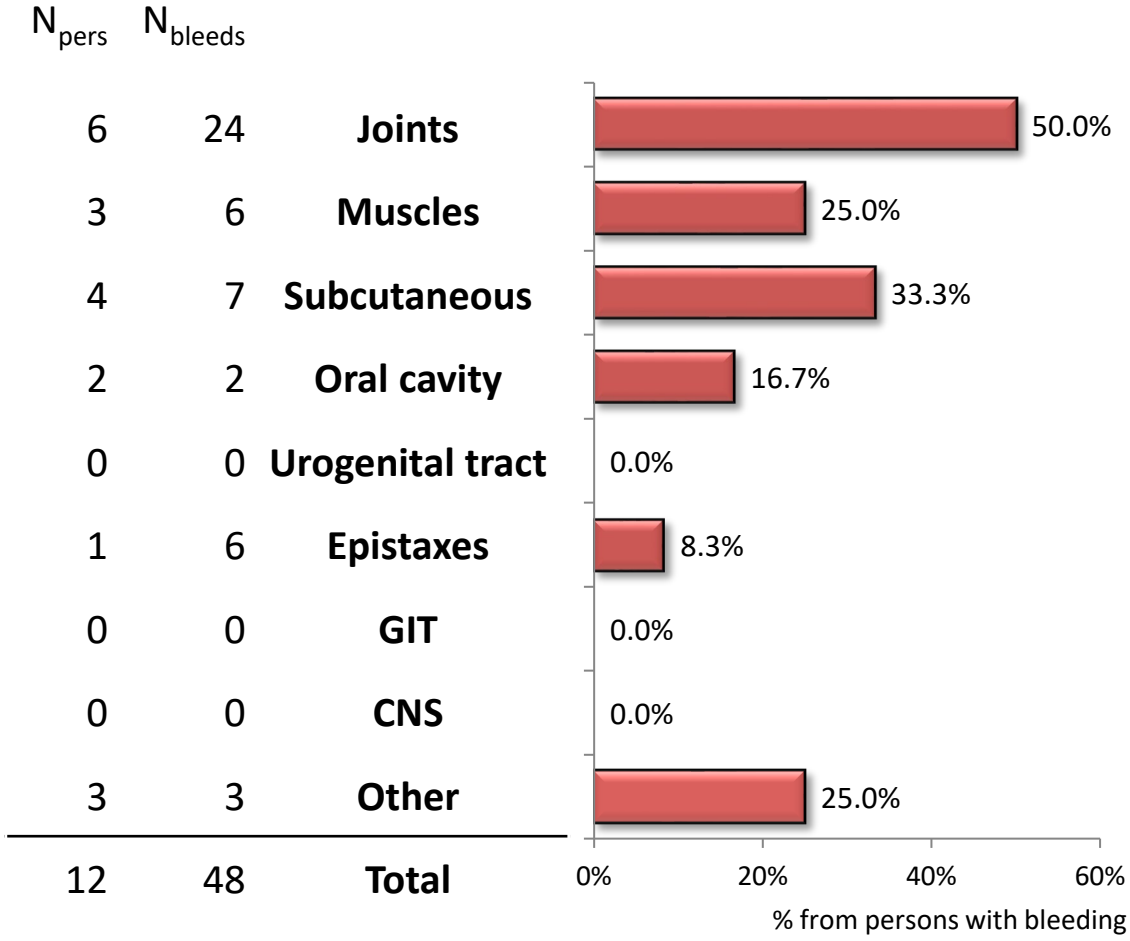
[1]Frequency of bleeding is missing in 2 adults.

Location of bleeds in 2022

Children
Haem B
N=38

12 (31.6%) children experienced bleeding at least once in year; 48 bleeds were recorded in total, 4 bleeds required hospitalization.
All of these 12 children have recorded location of their bleeds.

26 (68.4%) children recorded no bleed during year 2022.



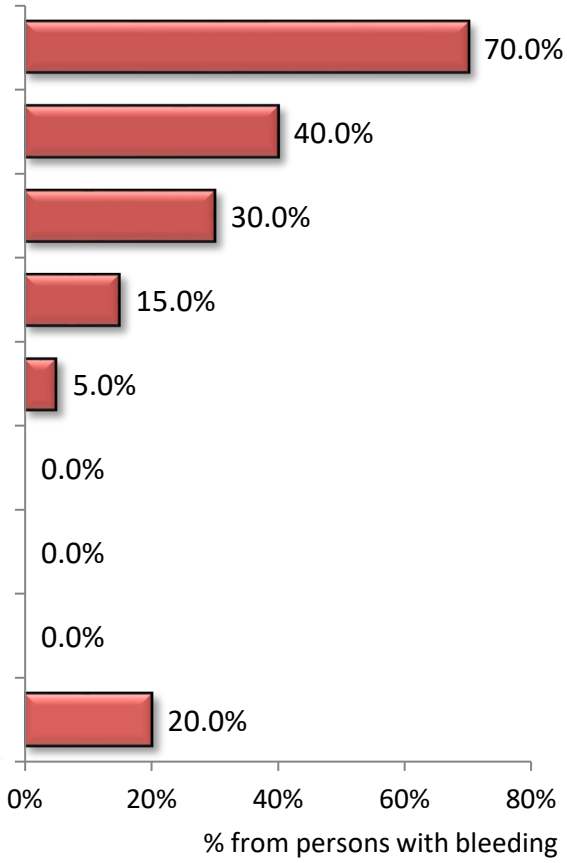
Location of bleeds in 2022

Adults
Haem B
N=63[1]

20 (31.6%) adults experienced bleeding requiring treatment at least once in year; 83 bleeds were recorded in total, 3 bleeds required hospitalization.
All of these 20 adults have recorded location of their bleeds.

43 (68.3%) adults have recorded no bleed during year 2022.

N_{pers}	N_{bleeds}		
14	57	**Joints**	70.0%
8	10	**Muscles**	40.0%
6	7	**Subcutaneous**	30.0%
3	4	**Oral cavity**	15.0%
1	1	**Urogenital tract**	5.0%
0	0	**Epistaxes**	0.0%
0	0	**GIT**	0.0%
0	0	**CNS**	0.0%
4	4	**Other**	20.0%
20	83	**Total**	

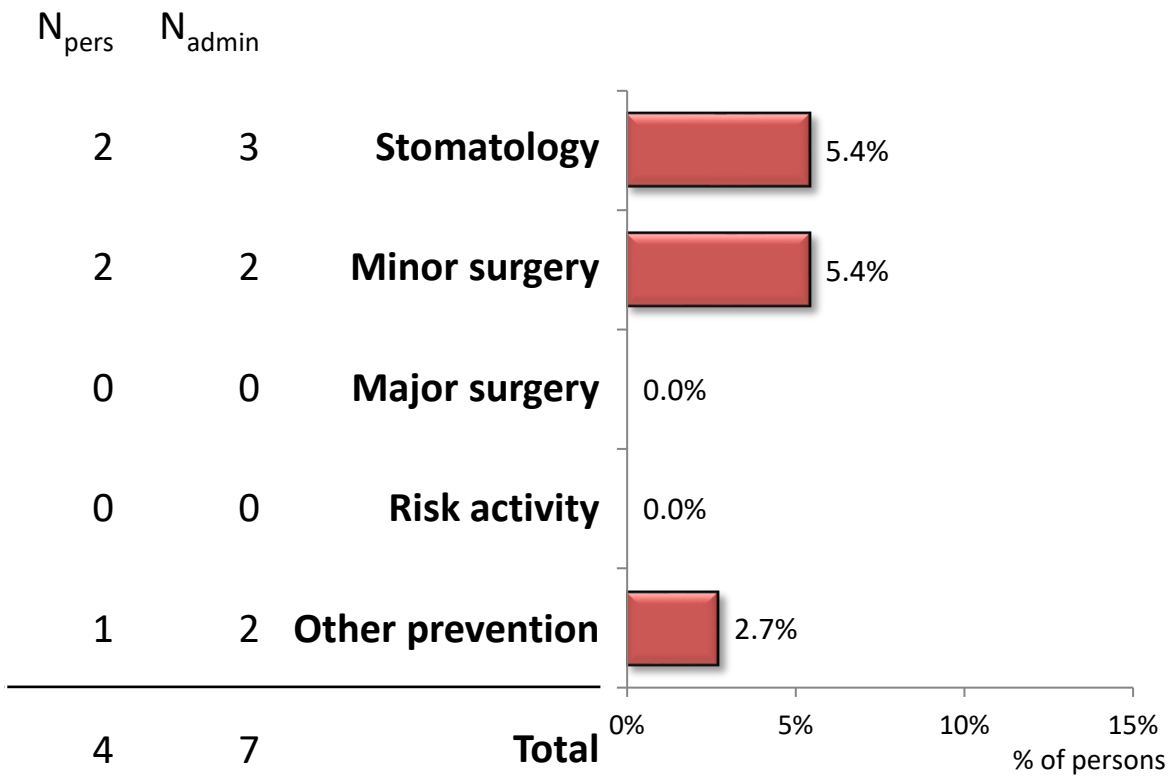


[1]*Frequency of bleeding is missing in 2 adults.*

Preventive administration in 2022

Children
Haem B
N=38

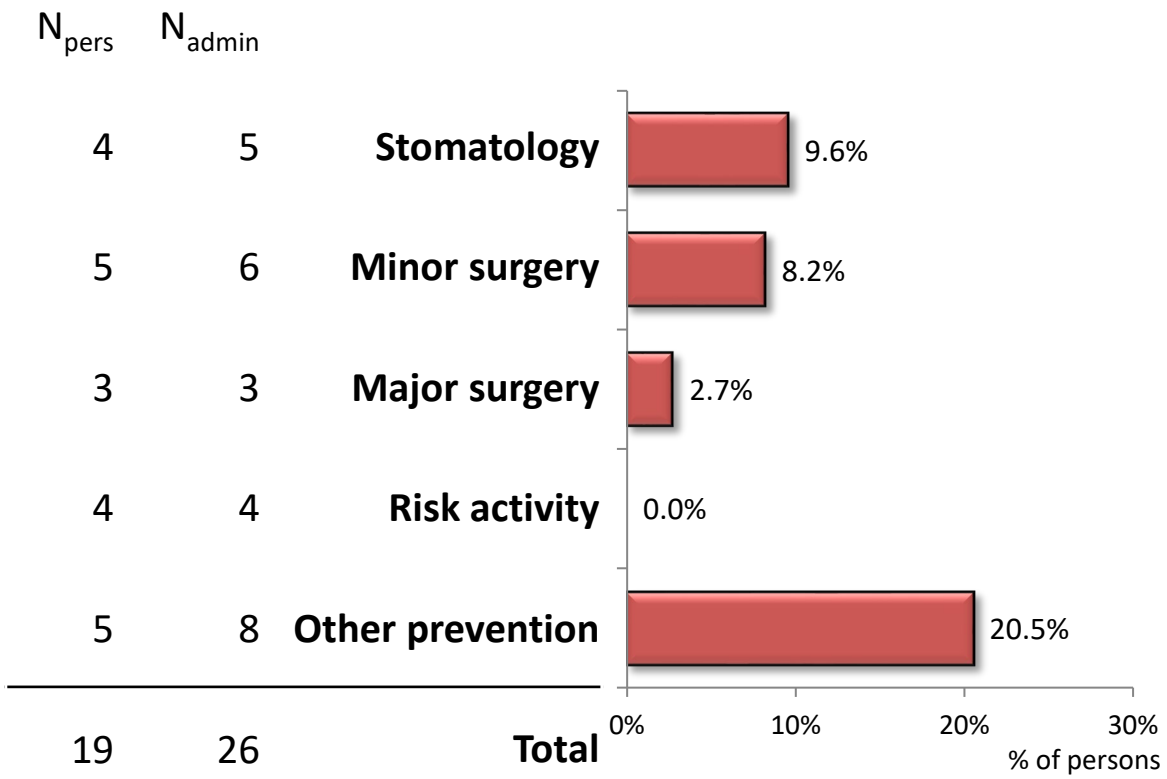
4 (10.5%) children were given factor to prevent bleeding during/before risk situation.
7 preventive administrations were recorded in total.



Preventive administration in 2022

Adults
Haem B
N=65

19 (29.2%) persons were given factor to prevent bleeding during/before risk situation.
26 preventive administrations were recorded in total.



ABR according to treatment regimen Haemophilia B without inhibitor

Annual bleeding rate according to treatment regimen

Children Haem B N=37*

Frequency of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*15*	*0*	*9*	*2*	*2*	*9*
Mean	*0.2*	*0.0*	*0.1*	*2.0*	***0.5***	***2.2***
Median (min – max)	*0 (0 – 1)*	*(–)*	*0 (0 – 1)*	*2 (0 – 4)*	***0.5 (0 – 1)***	***2 (0 – 9)***
Total no of recorded bleeds	*3*	*0*	*1*	*4*	***1***	***20***
Children on permanent prophylaxis	***0 (0%)***		***2 (18.2%)***		***9 (81.8%)***	
% of factor (FVIII) consumed by children on permanent prophylaxis	***0.0%***		***99.8%***		***99.9%***	

Location of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*15*	*0*	*9*	*2*	*2*	*9*
JOINT BLEEDS						
Mean	*0.0*	*0*	*0.1*	*2.0*	***0.0***	***0.8***
Median (range)	*0 (0 – 0)*	*(–)*	*0 (0 – 1)*	*2 (0 – 4)*	***0 (0 – 0)***	***0 (0 – 3)***
Total no of recorded bleeds	*0*	*0*	*1*	*4*	***0***	***7***
OTHER BLEEDS						
Mean	*0.2*	*0*	*0.0*	*0.0*	***0.5***	***1.6***
Median (range)	*0 (0 – 1)*	*(–)*	*0 (0 – 0)*	*0 (0 – 0)*	***0.5 (0 – 1)***	***0 (0 – 9)***
Total no of recorded bleeds	*3*	*0*	*0.00*	*0*	***1***	***14***

Treatment regimen:
<u>OD</u> = on demand and/or temporary prophylaxis
<u>prophy</u> = permanent prophylaxis

** without inhibitor*

Annual bleeding rate according to treatment regimen

Adults
Haem B
N=63*

Frequency of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*17*	*0*	*18*	*5*	*4*	*19*
Mean	*0.2*	*0.0*	*0.7*	*0.4*	***2.0***	***3.0***
Median (min – max)	*0 (0 – 2)*	*(–)*	*0 (0 – 4)*	*0 (0 – 2)*	***1 (0 – 6)***	***0 (0 – 18)***
Total no of recorded bleeds	*3*	*0*	*13*	*2*	***8***	***57***
Adults on permanent prophylaxis	***0 (0%)***		***5 (21.7%)***		***20 (80%)***	
% of factor (FVIII) consumed by children on permanent prophylaxis	***0.0%***		***70.8%***		***96.6%***	

Location of bleeding	Mild*		Moderate*		Severe*	
Treatment regimen	*OD*	*prophy*	*OD*	*prophy*	*OD*	*prophy*
N valid	*17*	*0*	*18*	*5*	*4*	*19*
JOINT BLEEDS						
Mean	*0.1*	*0*	*0.2*	*0.4*	***2.0***	***2.2***
Median (range)	*0 (0 – 1)*	*(–)*	*0 (0 – 3)*	*0 (0 – 2)*	***1.5 (0 – 5)***	***0 (0 – 14)***
Total no of recorded bleeds	*1*	*0*	*4*	*2*	***8***	***42***
OTHER BLEEDS						
Mean	*0.1*	*0*	*0.5*	*0.0*	***0.3***	***0.7***
Median (range)	*0 (0 – 1)*	*(–)*	*0 (0 – 4)*	*0 (0 – 0)*	***0 (0 – 1)***	***0 (0 – 4)***
Total no of recorded bleeds	*2*	*0*	*9*	*0*	***1***	***14***

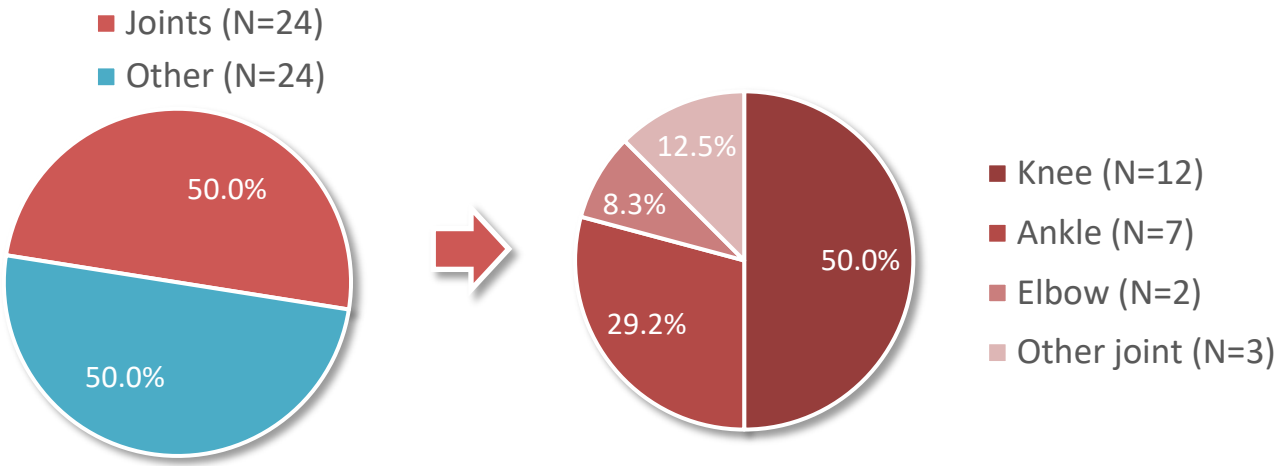
Treatment regimen:
<u>OD</u> = on demand and/or temporary prophylaxis
<u>prophy</u> = permanent prophylaxis

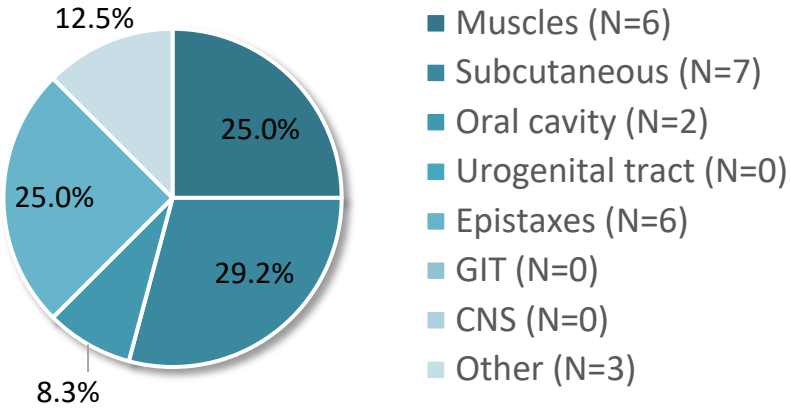
** without inhibitor; missing frequency of bleeding in 2 adults*

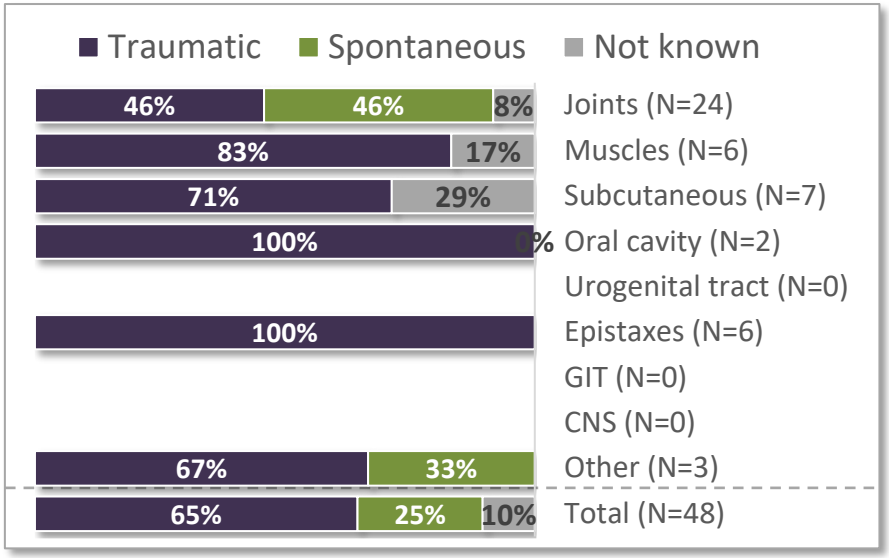
Location and etiology of bleeds

Children Haem B N=48*

** number of bleeds*







Detailed treatment of bleeds

Children Haem B N=48*

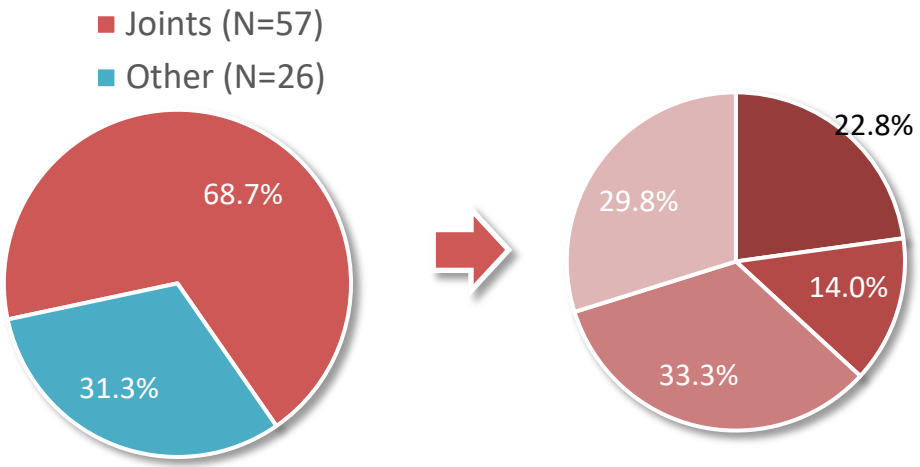
** number of bleeds*

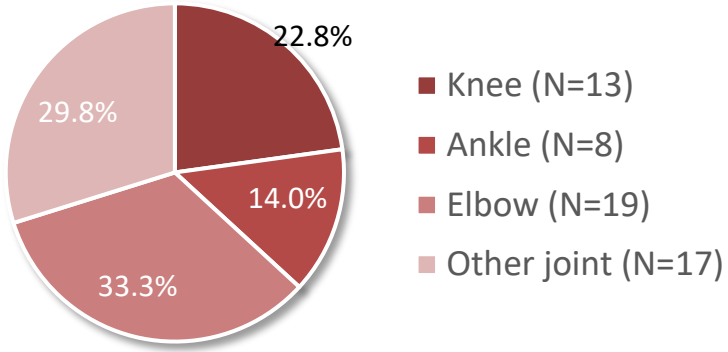
	Joints	Muscles	Subcuta-neous	Oral cavity	Urogenital tract	Epistaxes	GIT	CNS	Other	Total
No. of bleeds	*24*	*6*	*7*	*2*	*0*	*6*	*0*	*0*	*3*	***48***
FIX consumption per bleed (IU), valid N	*10*	*2*	*5*	*2*		*2*			*2*	***23***
geometric mean	3270.4	11225.0	715.5	1000.0		500.0			500.0	**1702.5**
median	**3500.0**	**13500.0**	**500.0**	**1250.0**		**500.0**			**500.0**	**2000.0**
min – max	500–9000	6000–21000	500–3000	500–2000		500–500			500–500	**500–21000**
sum	41 000	27 000	5 000	2 500		1 000			1 000	**77 500**
No. of doses per bleed										
geometric mean	1.7	2.7	1.0	1.4		1.0			3.4	**1.7**
median	**1**	**2**	**1**	**2**		**0**			**3**	**1**
min – max	0–4	0–7	0–1	1–2		0–1			1–13	**0–13**
Duration of therapy per bleed, days										
geometric mean	1.9	3.1	1.0	2.4		1.0			3.3	**1.9**
median	**2**	**2**	**1**	**3**		**1**			**6**	**2**
min – max	1–14	2–12	1–1	2–3		1–1			1–6	**1–14**
N (%) with hospitalization	*1 (4.2%)*	*1 (16.7%)*	*0 (0%)*	*1 (50%)*		*0 (0%)*			*1 (33.3%)*	***4 (8.3%)***
N (%) with rebleeding	*6 (25%)*	*2 (33.3%)*	*2 (28.6%)*	*0 (0%)*		*4 (66.7%)*			*0 (0%)*	***14 (29.2%)***

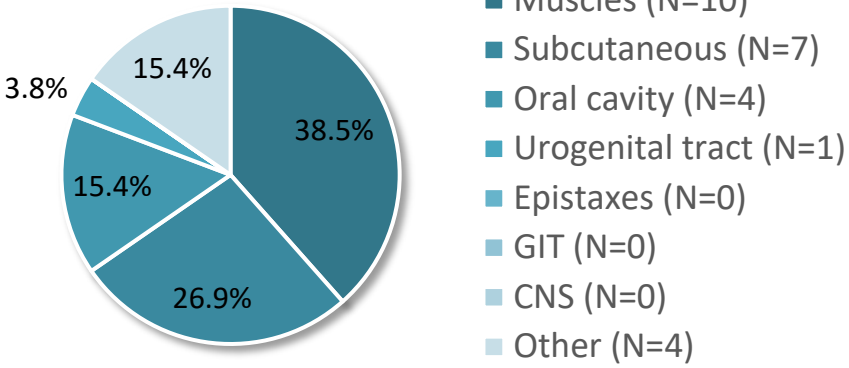
Location and etiology of bleeds

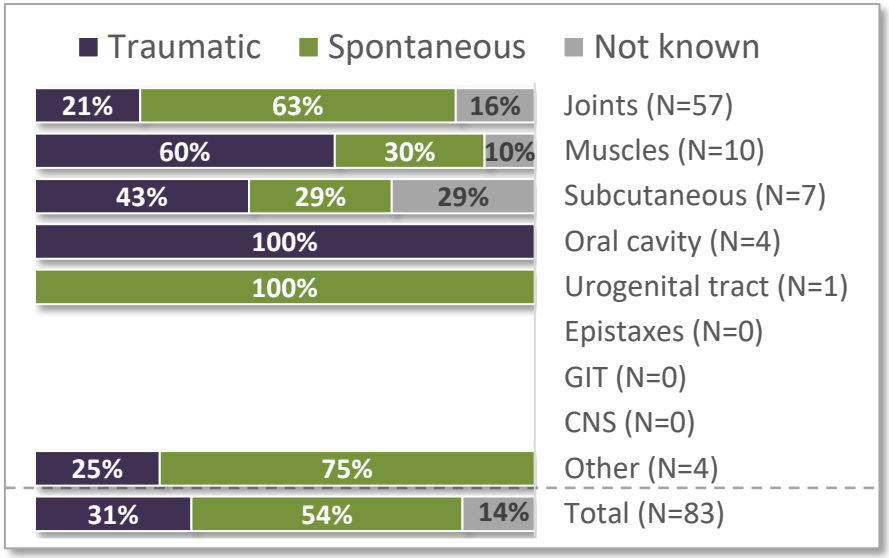
Adults
Haem B
N=83*

** number of bleeds*









Detailed treatment of bleeds

Adults
Haem B
N=83*

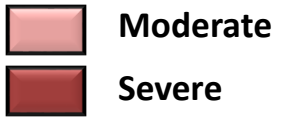
** number of bleeds*

	Joints	Muscles	Subcuta-neous	Oral cavity	Urogenital tract	Epistaxes	GIT	CNS	Other	Total
No. of bleeds	*57*	*10*	*7*	*4*	*1*	*0*	*0*	*0*	*4*	***83***
FIX consumption per bleed (IU), valid N	*57*	*10*	*7*	*4*	*1*				*4*	***83***
geometric mean	4169.0	6274.9	6055.6	3191.0	13200.0				6447.4	**4620.2**
median	**4000.0**	**6000.0**	**6000.0**	**4200.0**	**13200.0**				**6000.0**	**6000.0**
min – max	600–18000	1200–49500	2000–18000	1000–7200	13200–13200				6000–8000	**600–49500**
sum	313 800	103 500	53 600	16 600	13 200				26 000	**526 700**
No. of doses per bleed										
geometric mean	1.4	2.5	1.6	2.5	14.0				1.5	**1.6**
median	**1**	**3**	**1**	**3**	**14**				**1**	**1**
min – max	1–5	1–16	1–5	1–7	14–14				1–5	**1–16**
Duration of therapy per bleed, days										
geometric mean	1.3	2.4	1.6	2.2	4.0				2.1	**1.5**
median	**1**	**2**	**1**	**3**	**4**				**1**	**1**
min – max	1–7	1–12	1–5	1–4	4–4				1–19	**1–19**
N (%) with hospitalization	*2 (3.5%)*	*1 (10%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*				*0 (0%)*	***3 (3.6%)***
N (%) with rebleeding	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*	*0 (0%)*				*0 (0%)*	***0 (0%)***

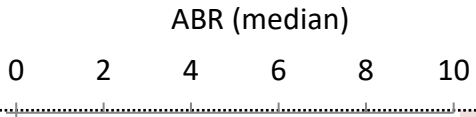
ABR according to centres Haemophilia B (PWHB)

Annual bleeding rate on permanent prophylaxis

HaemB on prophy
Paed. centres
N=11



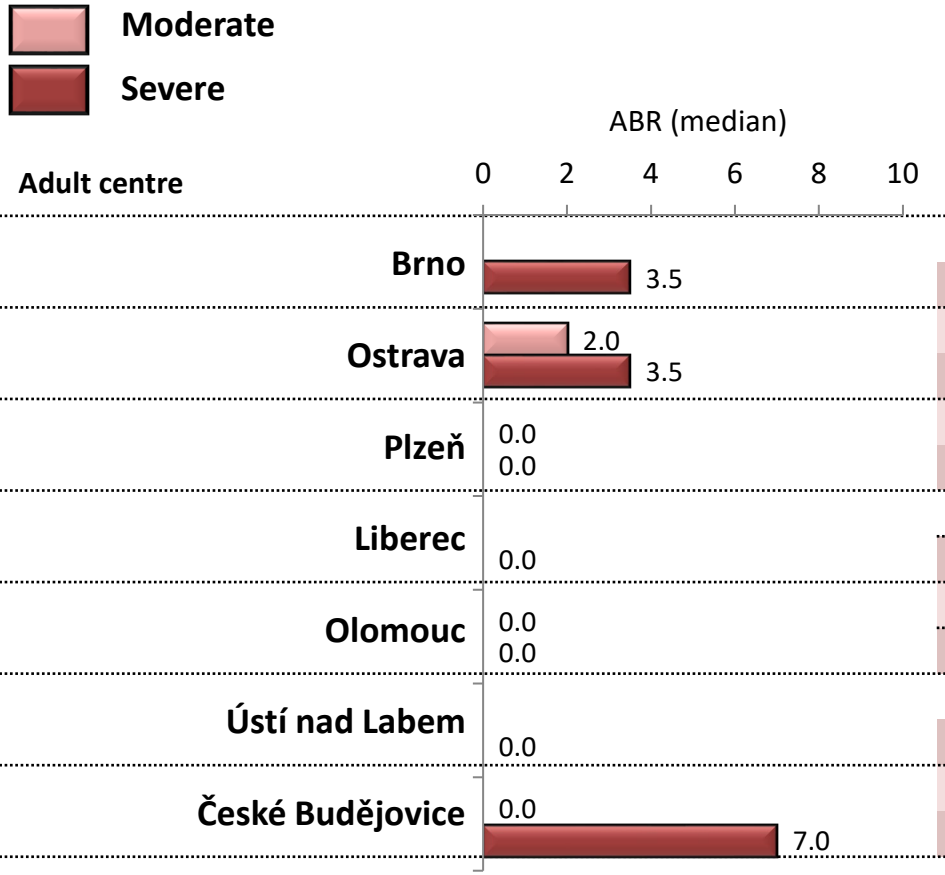
Frequency of bleeding in PWHB without inhibitor on permanent prophylaxis



Paediatric centre	N	Mean	Median	Min	Max	Severity
Praha	2	2.0	2.0	0	4	Moderate
	4	3.5	2.5	0	9	Severe
Brno	0					
	1	2.0	2.0	2	2	Severe
Ostrava	0					
	3	0.0	0.0	0	0	Severe
Ústí nad Labem	0					
	1	4.0	4.0	4	4	Severe

Annual bleeding rate on permanent prophylaxis

HaemB on prophy
Adult centres
N=17



Frequency of bleeding in PWHB without inhibitor on permanent prophylaxis

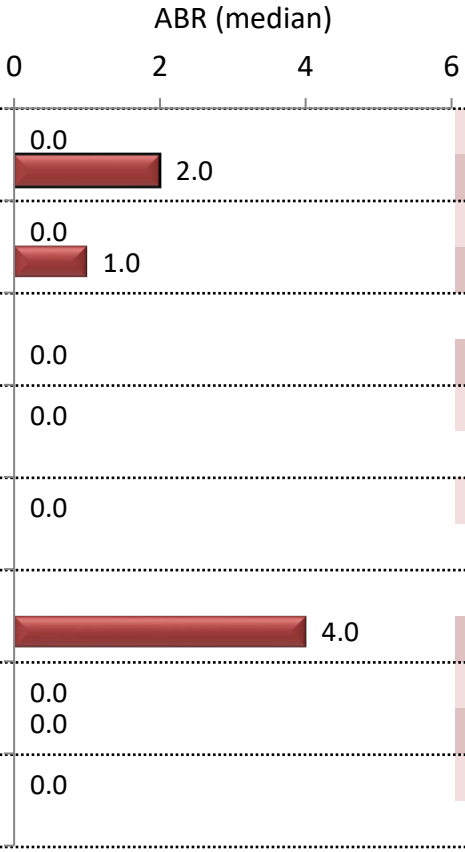
Adult centre	N	Mean	Median	Min	Max	Severity
Brno	0					
	6	4.0	3.5	0	11	Severe
Ostrava	1	2.0	2.0	2	2	Moderate
	4	6.5	3.5	1	18	Severe
Plzeň	2	0.0	0.0	0	0	Moderate
	4	0.0	0.0	0	0	Severe
Liberec	0					
	1	0.0	0.0	0	0	Severe
Olomouc	1	0.0	0.0	0	0	Moderate
	1	0.0	0.0	0	0	Severe
Ústí nad Labem	0					
	1	0.0	0.0	0	0	Severe
České Budějovice	1	0.0	0.0	0	0	Moderate
	1	7.0	7.0	7	7	Severe

Annual bleeding rate regardless prophylaxis

HaemB
Paed. centres
N=23

Moderate
Severe

Frequency of bleeding in PWHB without inhibitor regardless of prophylaxis



Paediatric centre	N	Mean	Median	Min	Max	% on permanent prophylaxis
Praha	6	0.8	0.0	0	4	33.3%
	5	3.0	2.0	0	9	80.0%
Brno	1	0.0	0.0	0	0	0.0%
	2	1.0	1.0	0	2	50.0%
Ostrava	0					
	3	0.0	0.0	0	0	100.0%
České Budějovice	1	0.0	0.0	0	0	0.0%
	0					
Hradec Králové	1	0.0	0.0	0	0	0.0%
	0					
Ústí nad Labem	0					
	1	4.0	4.0	4	4	100.0%
Plzeň	1	0.0	0.0	0	0	0.0%
	1	0.0	0.0	0	0	100.0%
Olomouc	1	0.0	0.0	0	0	0.0%
	0					

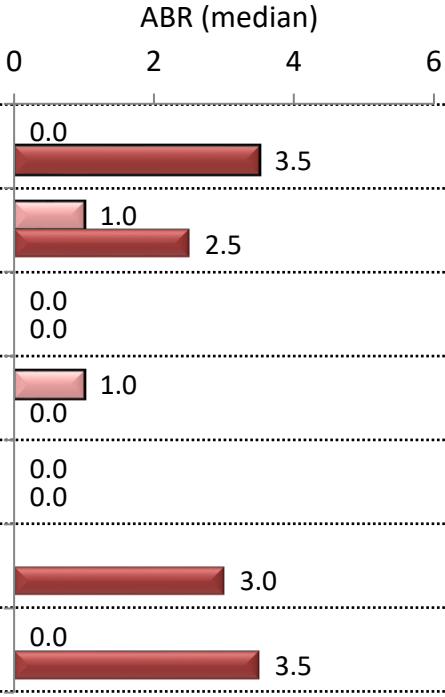
Annual bleeding rate regardless prophylaxis

HaemB
Adult centres
N=45*

** missing ABR in 2 adults*

Moderate
Severe

Frequency of bleeding in PWHB without inhibitor regardless of prophylaxis



Adult centre	N*	Mean	Median	Min	Max	% on permanent prophylaxis
Brno	5	0.2	0.0	0	1	0.0%
	6	4.0	3.5	0	11	100.0%
Ostrava	3	1.0	1.0	0	2	33.3%
	6	4.7	2.5	0	18	66.7%
Plzeň	3	0.0	0.0	0	0	66.7%
	4	0.0	0.0	0	0	100.0%
Liberec	2	1.0	1.0	0	2	0.0%
	1	0.0	0.0	0	0	100.0%
Olomouc	8	1.1	0.0	0	4	12.5%
	1	0.0	0.0	0	0	66.7%
Ústí nad Labem	0					
	2	3.0	3.0	0	6	50.0%
České Budějovice	2	0.0	0.0	0	0	50.0%
	5	0.2	0.0	0	1	0.0%

Prophylactic regimens and treatment outcomes

HaemB
Paed. centres
N=23

Paediatric centre	Severity	Total N	PERMANENT PROPHYLAXIS								ON-DEMAND / TEMPORARY PROPHY		
			% of patients	N	Dosing of **SHL** prophylaxis (IU/kg per week)		Dosing of **EHL** prophylaxis (IU/kg per week)		ABR		N	ABR	
					Mean	Median	Mean	Median	Mean	Median		Mean	Median
Praha	**Moderate**	6	**33.3%**	2			29.4	**29.4**	2.0	**2.0**	4	0.3	**0.0**
	Severe	5	**80.0%**	4			42.1	**34.5**	3.5	**2.5**	1	1.0	**1.0**
Brno	**Moderate**	1	**0.0%**	0							1	0.0	**0.0**
	Severe	2	**50.0%**	1			45.5	**45.5**	2.0	**2.0**	1	0.0	**0.0**
Ostrava	**Moderate**	0	**0.0%**	0							0		
	Severe	3	**100.0%**	3	72.0	**72.0**	20.0	**20.0**	0.0	**0.0**	0		
Č. Budějovice	**Moderate**	1	**0.0%**	0							1	0.0	**0.0**
	Severe	0	**0.0%**	0							0		
Hradec Králové	**Moderate**	1	**0.0%**	0							1	0.0	**0.0**
	Severe	0	**0.0%**	0							0		
Ústí nad Labem	**Moderate**	0	**0.0%**	0							0		
	Severe	1	**100.0%**	1					4.0	**4.0**	0		
Plzeň	**Moderate**	1	**0.0%**	0							1	0.0	**0.0**
	Severe	1	**100.0%**	1			58.8	**58.8**	0.0	**0.0**	0		
Olomouc	**Moderate**	1	**0.0%**	0							1	0.0	**0.0**
	Severe	0	**0.0%**	0							0		

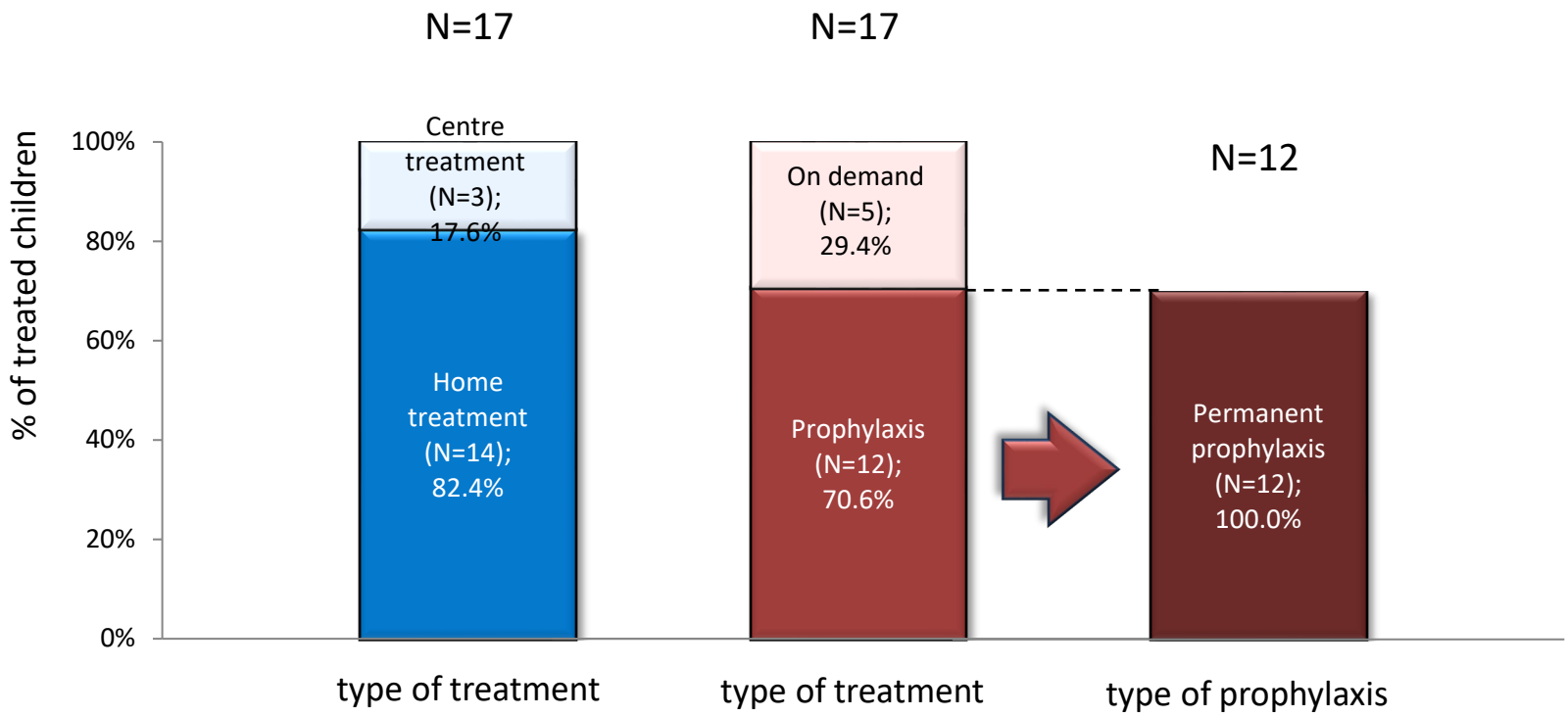
Prophylactic regimens and treatment outcomes

HaemB
Adult centres
N=47

Adult centre	Severity	Total N	PERMANENT PROPHYLAXIS									ON-DEMAND / TEMPORARY PROPHY			
			% of patients	N	Dosing of **SHL** prophylaxis (IU/kg per week)		Dosing of **EHL** prophylaxis (IU/kg per week)		ABR		Age	N	ABR		Age
					Mean	Median	Mean	Median	Mean	Median	Median		Mean	Median	Median
Brno	**Moderate**	5	**0.0%**	0								5	0.2	**0.0**	50
	Severe	6	**100.0%**	6			47.0	**49.5**	4.0	**3.5**	33	0			
Ostrava	**Moderate**	3	**33.3%**	1			28.2	**28.2**	2.0	**2.0**	23	2	0.5	**0.5**	61
	Severe	6	**66.7%**	4	46.9	**52.2**	55.0	**55.0**	6.5	**3.5**	59	2	1.0	**1.0**	52
Plzeň	**Moderate**	3	**66.7%**	2			23.5	23.5	0.0	**0.0**	56	1	0.0	**0.0**	60
	Severe	4	**100.0%**	4	56.6	**56.6**	21.8	**21.1**	0.0	**0.0**	43	0			
Liberec	**Moderate**	2	**0.0%**	0								2	1.0	**1.0**	34
	Severe	1	**100.0%**	1			55.3	**55.3**	0.0	**0.0**	30	0			
Olomouc	**Moderate**	8	**12.5%**	1	40.8	**40.8**	20.4	20.4	0.0	**0.0**	43	7	1.3	**0.0**	54
	Severe	3	**66.7%**	2			35.2	**35.2**	0.0	**0.0**	42	1	0.0	**0.0**	56
Ústí n. Labem	**Moderate**	0	**0.0%**	0								0			
	Severe	2	**50.0%**	1			19.2	**19.2**	0.0	**0.0**	27	1	6.0	**6.0**	51
Č. Budějovice	**Moderate**	2	**50.0%**	1			23.1	23.1	0.0	**0.0**	56	1	0.0	**0.0**	35
	Severe	2	**50.0%**	1			6.5	**6.5**	7.0	**7.0**	49	1	0.0	**0.0**	59

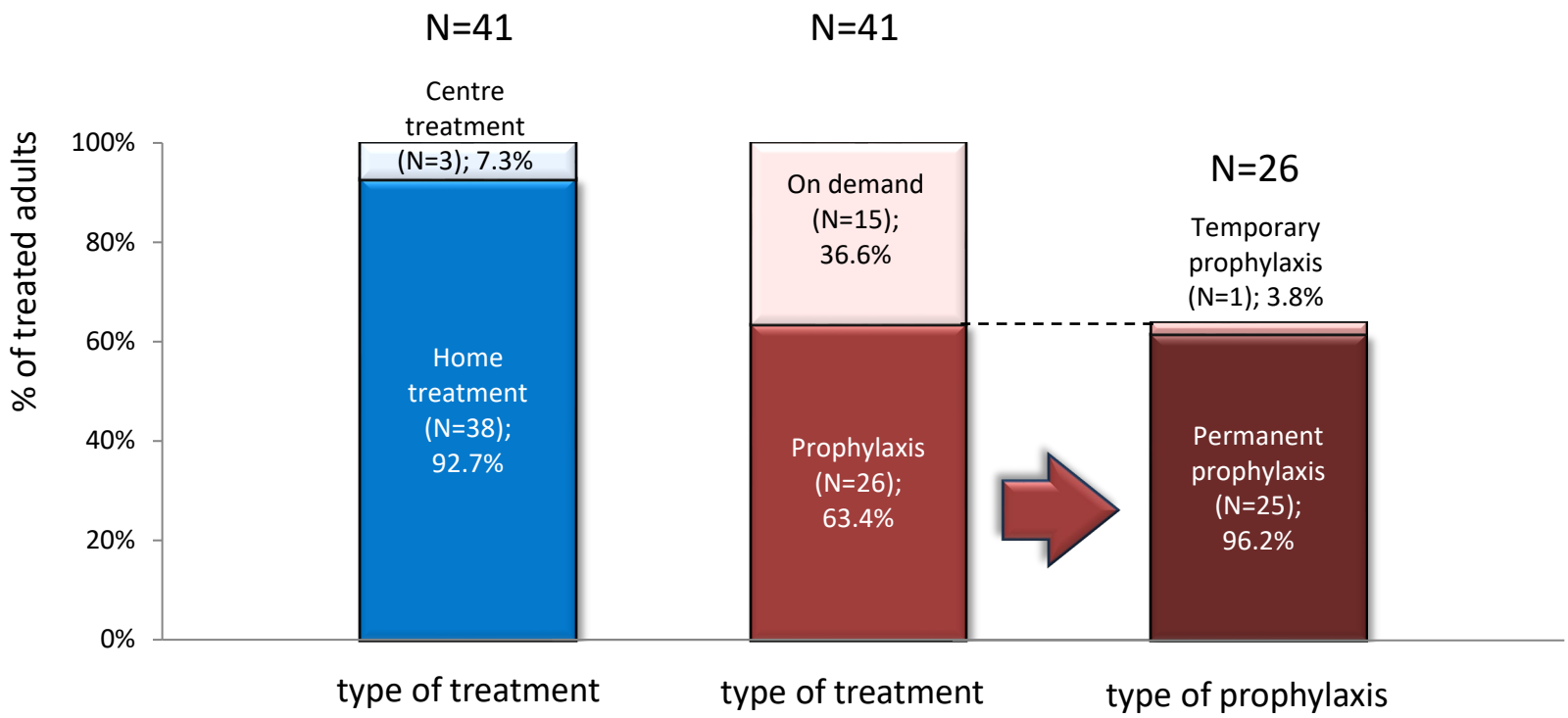
Children
Haem B
N=17

Type of treatment (subgroup of treated patients)



Adults
Haem B
N=41

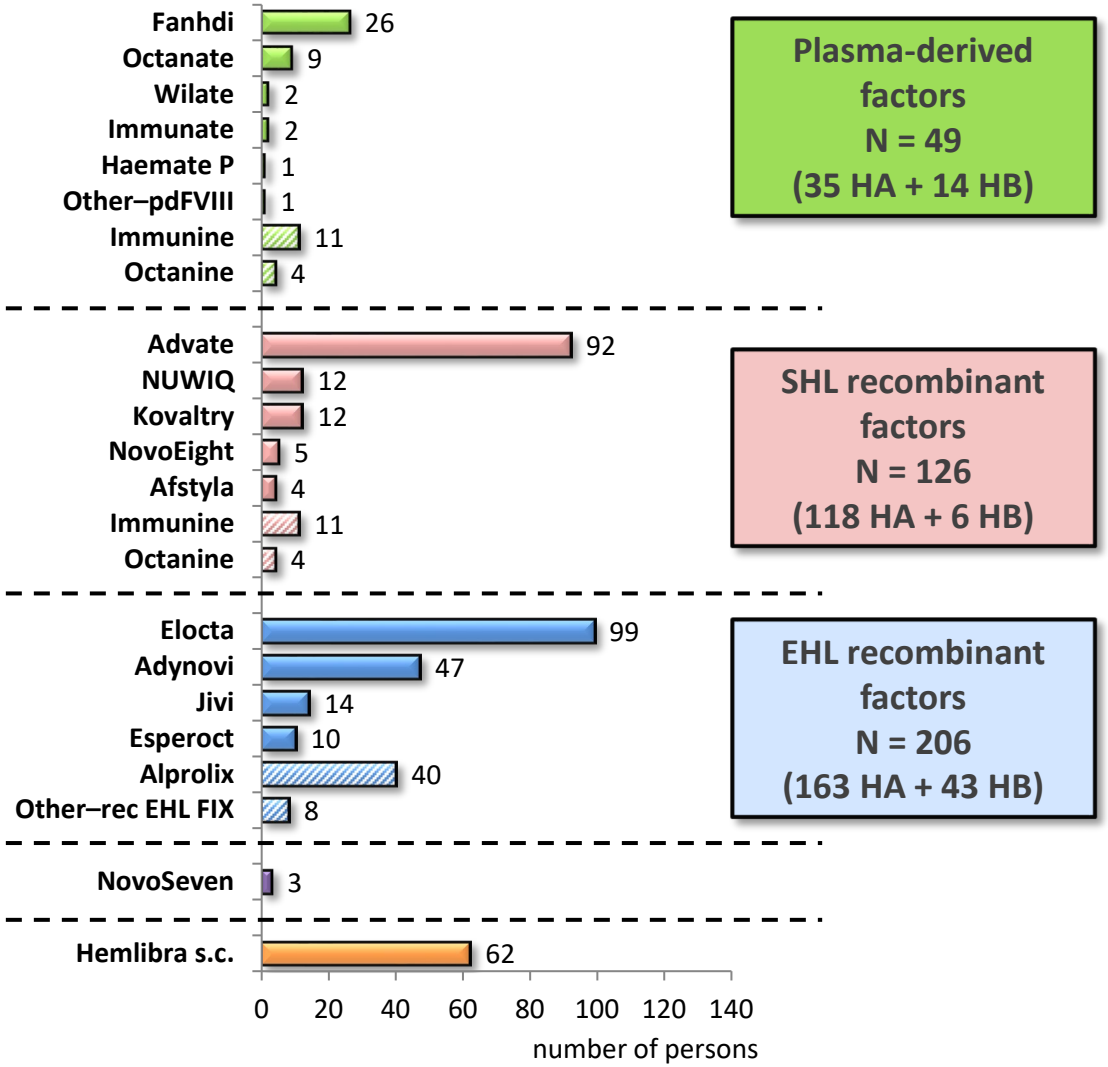
Type of treatment (subgroup of treated patients)



Treatment data and factor consumption Haemophilia A and B

Treatment

All
N=394[1]



Plasma-derived factors
N = 49
(35 HA + 14 HB)

SHL recombinant factors
N = 126
(118 HA + 6 HB)

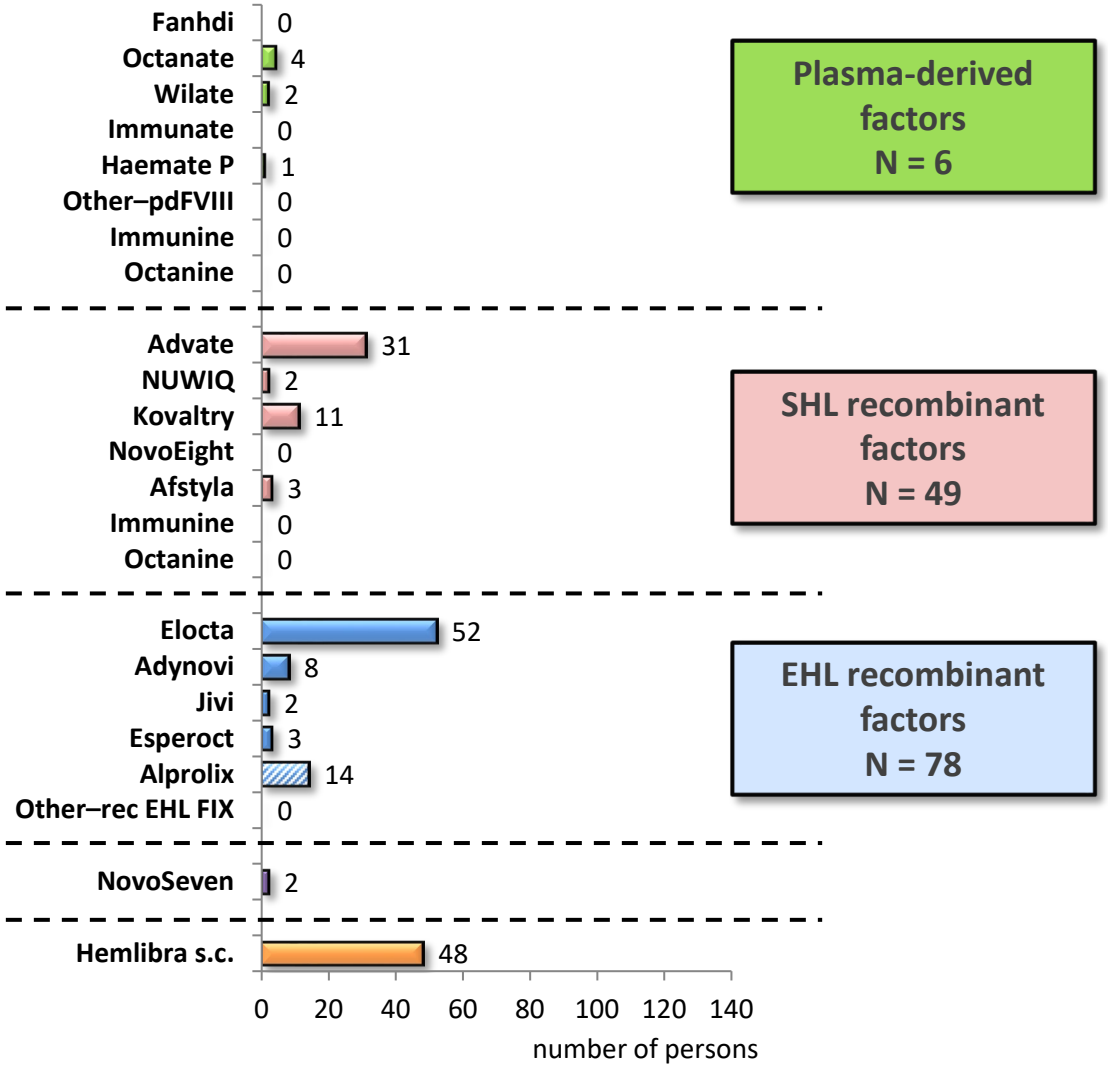
EHL recombinant factors
N = 206
(163 HA + 43 HB)

394 persons (52% of all PWH) were treated in 2022 (175 persons received standard factor concentrates, 206 persons received EHL factors, 1 by-pass therapy and 62 emicizumab, in 4 persons data are not available; 54 persons received more than one type/brand of concentrate).
9 persons were treated with both plasma-derived and recombinant factor.

[1]missing type of treatment in 4 adults

Treatment

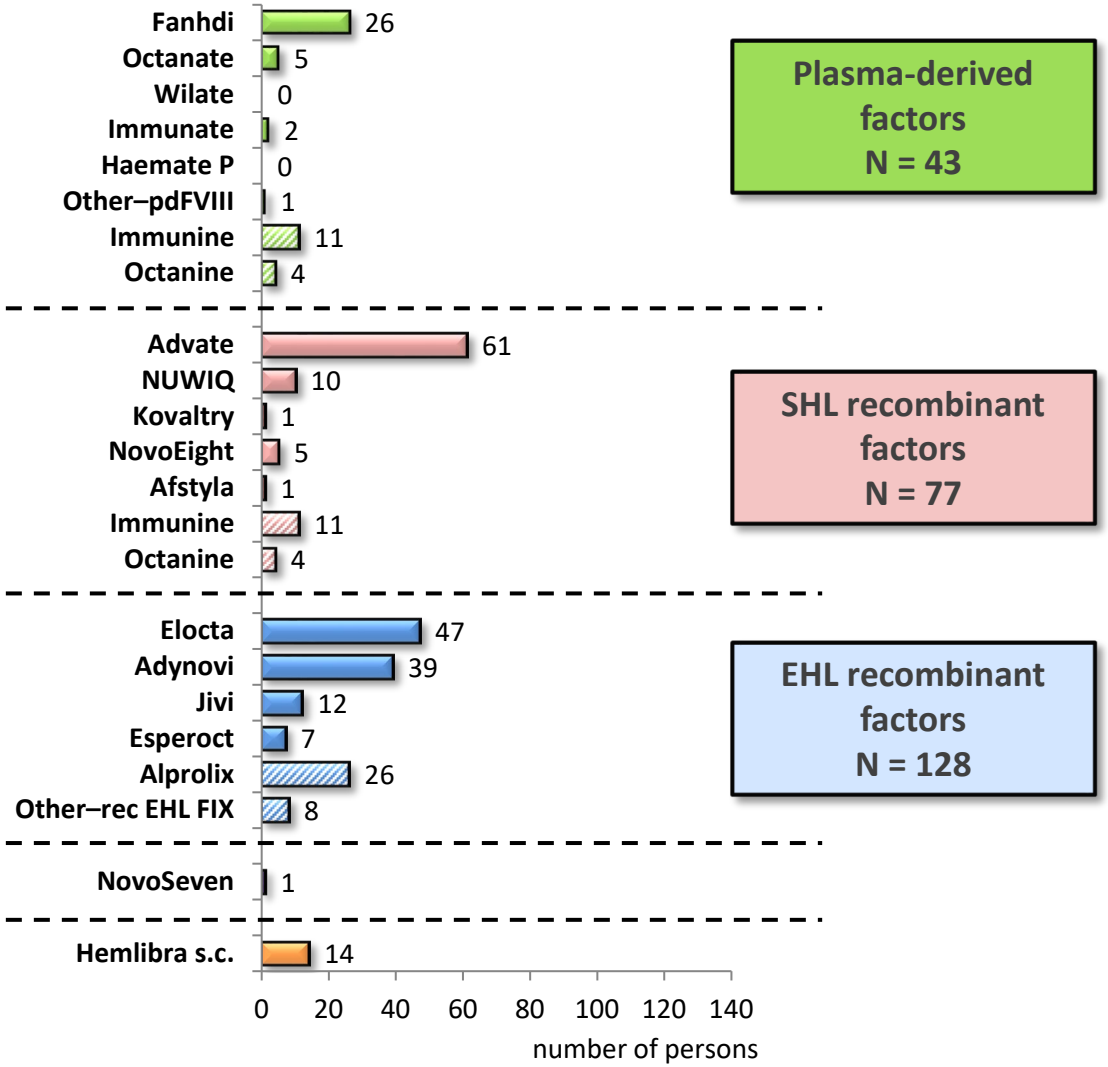
Children
N=152



152 children (53.1% of all PWH) were treated in 2022 (55 children received standard factor concentrates, 78 EHL factors, 2 by-pass therapy and 48 children emicizumab; 27 children received more than one type/brand of concentrate). None child were treated with both plasma-derived and recombinant factor.

Treatment

Adults
N=242[1]



242 adults (51.4% of all PWH) were treated in 2022 (120 adults received standard factor concentrates, 128 EHL factors, 1 by-pass therapy adn 14 adults emicizumab; 43 adults received more than one type/brand of concentrate).
9 adults were treated with both plasma-derived and recombinant factor.

[1]*missing type of treatment in 4 adults*

Comparison of treatment in years 2022 and 2021

	2022			2021		
	N	% of all PWHs	% treated PWHs	N	% of all PWHs	% treated PWHs
All persons treated with factor concentrates*	394	52.0	100.0	397	48.5	100.0
Plasma-derived factor	*49*	*6.5*	*12.4*	*73*	*8.9*	*18.4*
Recombinant factor	*126*	*16.6*	*32.0*	*200*	*24.4*	*50.4*
Recombinant f. EHL	*206*	*27.2*	*52.3*	*200*	*24.4*	*50.4*
Emicizumab	*62*	*8.2*	*15.7*	*39*	*4.8*	*9.8*
Without treatment	363	48.0	-	421	51.5	-
Total	757	100.0	-	818	100.0	-

** One patient could have more type of factor concentrates.*

Children

Comparison of treatment in years 2022 and 2021

	2022			2021		
	N	% of all PWHs	% treated PWHs	N	% of all PWHs	% treated PWHs
All persons treated with factor concentrates*	152	53.1	100.0	122	44.4	100.0
Plasma-derived factor	*6*	*2.1*	*3.9*	*4*	*1.5*	*3.3*
Recombinant factor	*49*	*17.1*	*32.2*	*63*	*22.9*	*51.6*
Recombinant f. EHL	*78*	*27.3*	*51.3*	*72*	*26.2*	*59.0*
Emicizumab	*48*	*16.8*	*31.6*	*28*	*10.2*	*23*
Without treatment	134	46.9	-	153	55.6	-
Total	286	100.0	-	275	100.0	-

** One patient could have more type of factor concentrates.*

Adults

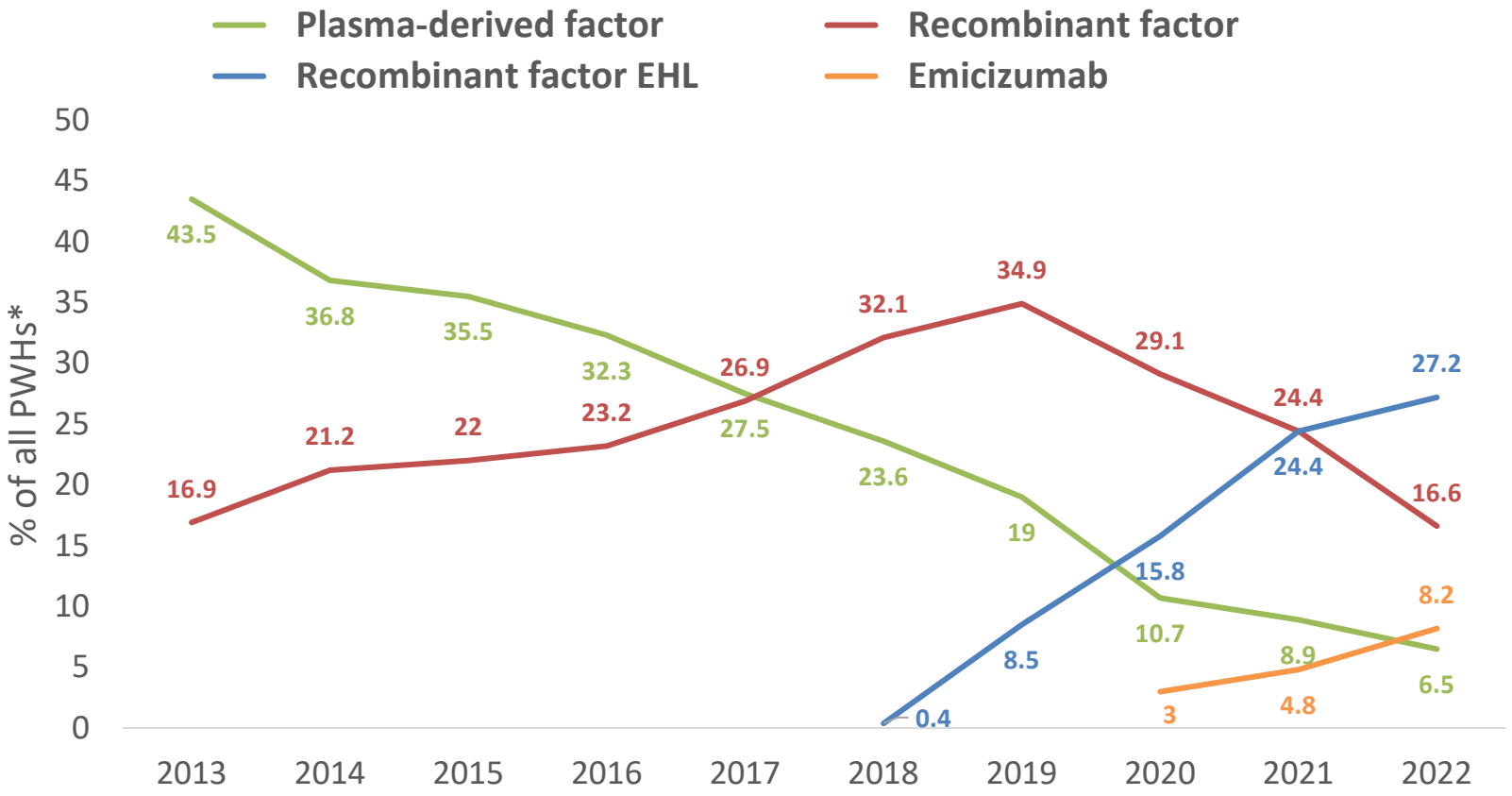
Comparison of treatment in years 2022 and 2021

	2022			2021		
	N	% of all PWHs	% treated PWHs	N	% of all PWHs	% treated PWHs
All persons treated with factor concentrates*	242	51.4	100.0	275	50.6	100.0
Plasma-derived factor	*43*	*9.1*	*17.8*	*69*	*12.7*	*25.1*
Recombinant factor	*77*	*16.3*	*31.8*	*137*	*25.2*	*49.8*
Recombinant f. EHL	*128*	*27.2*	*52.9*	*128*	*23.6*	*46.5*
Emicizumab	*14*	*3.0*	*5.8*	*11*	*2*	*4*
Without treatment	229	48.6	-	268	49.4	-
Total	471	100.0	-	543	100.0	-

** One patient could have more type of factor concentrates.*

All

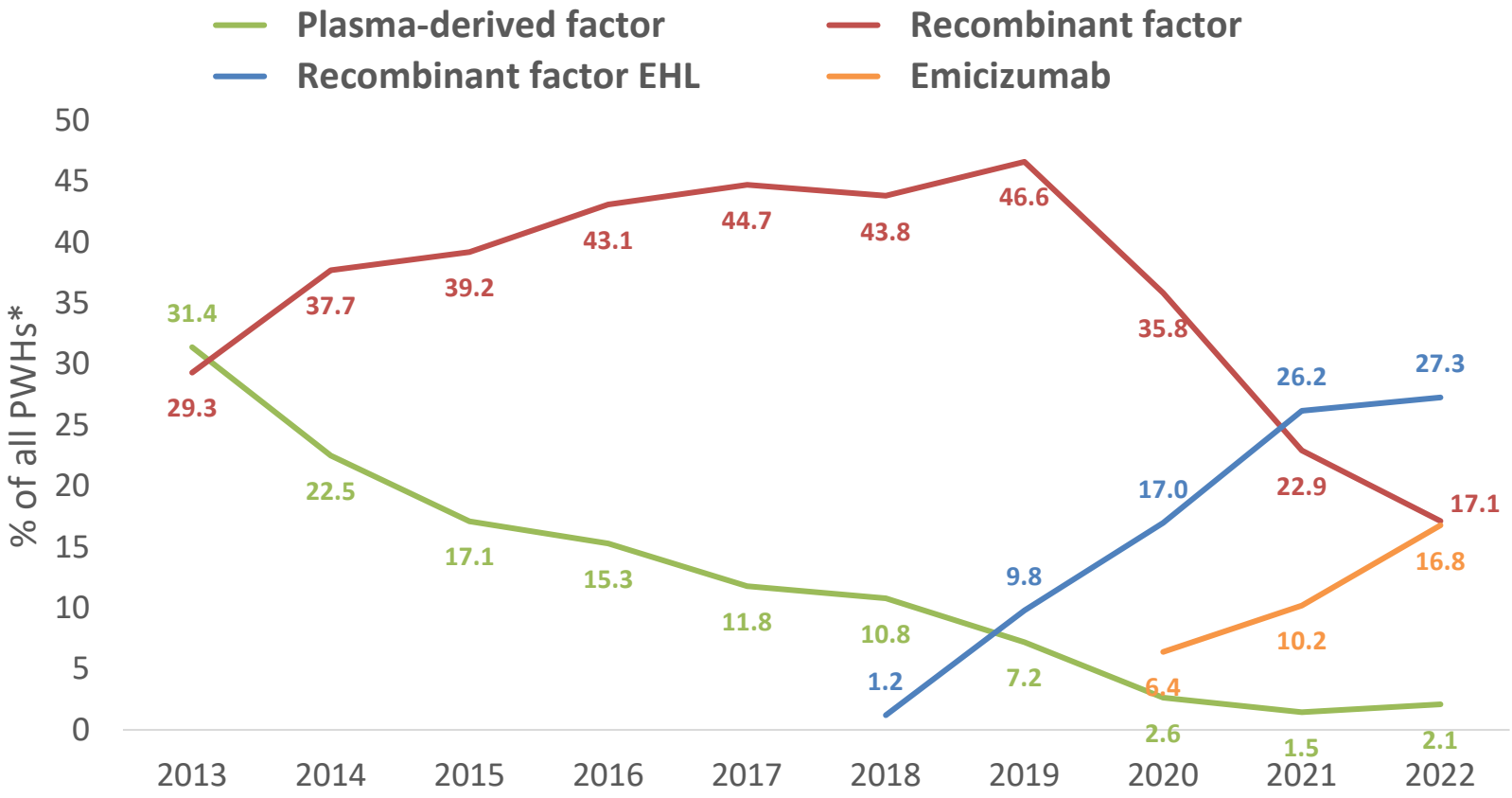
Comparison of treatment in years



** One patient could have more type of factor concentrates.*

Children

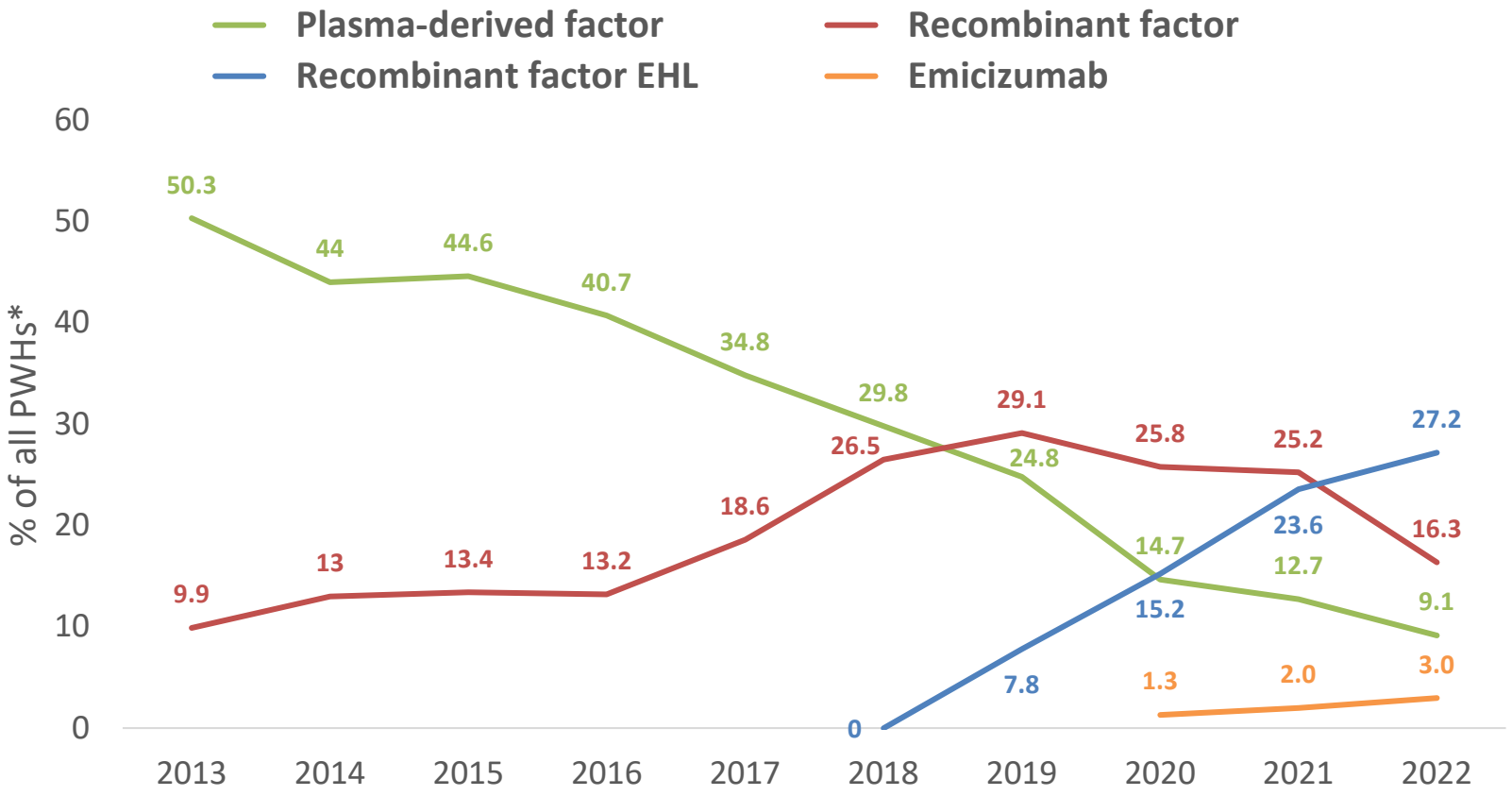
Comparison of treatment in years



** One patient could have more type of factor concentrates.*

Comparison of treatment in years

Adults



** One patient could have more type of factor concentrates.*

Consumption of drugs

All

	Drug (IU)	Total annual consumption	Number of treated persons	Average annual consumption per treated person
FVIII (IU)	Fanhdi	1 225 900	26	47 150.0
	Octanate	1 001 750	9	111 305.6
	Wilate	13 000	2	6 500.0
	Immunate	1 000	2	500.0
	Haemate P	6 000	1	6 000.0
	Other plasma-derived	300	1	300.0
	PD FVIII total	**2 247 650**	**34**	**66 107.4**
	Advate	6 961 000	92	75 663.0
	NUWIQ	2 709 500	12	225 791.7
	Kovaltry	209 750	12	17 479.2
	NovoEight	986 500	5	197 300.0
	Afstyla	316 500	4	79 125.0
	SHL REC FVIII total	**10 477 250**	**116**	**90 321.1**
	Standard FVIII total	*12 724 900*	*148*	*85 979.1*
	Elocta	14 541 574	99	146 884.6
	Adynovi	11 415 700	47	242 887.2
	Jivi	3 327 000	14	237 642.9
	Esperoct	2 131 200	10	213 120.0
	EHL REC FVIII total	*31 139 474*	*163*	*191 039.7*
	FVIII total	**43 864 374**	**295**	**148 692.8**
FIX (IU)	Immunine	304 200	11	27 654.5
	Octanine	299 500	4	74 875.0
	FIX PD total	**603 700**	**14**	**43 121.4**
	Rixubis	589 500	6	98 250.0
	Benefix	10 000	1	10 000.0
	FIX REC total	**599 500**	**7**	**85 642.9**
	Standard FIX total	*1 203 200*	*21*	*57 295.2*
	Alprolix	3 672 870	40	91 821.8
	Other–rec EHL FIX	471 600	8	58 950.0
	EHL REC FIX total	*4 144 470*	*43*	*96 383.0*
	FIX total	**5 347 670**	**56**	**95 494.1**
By-pass	NovoSeven (mg)	1 378.0	3	459.3
Emicizumab	Hemlibra s.c. (mg)	165 006	62	2 661.4

Consumption of drugs

Children

	Drug (IU)	Total annual consumption	Number of treated persons	Average annual consumption per treated person
FVIII (IU)	Fanhdi	0	0	
	Octanate	778 750	4	194 687.5
	Wilate	13 000	2	6 500.0
	Immunate	0	0	
	Haemate P	6 000	1	6 000.0
	Other plasma-derived	0	0	
	PD FVIII total	**797 750**	**6**	**132 958.3**
	Advate	1 143 750	31	36 895.2
	NUWIQ	1 030 000	2	515 000.0
	Kovaltry	181 750	11	16 522.7
	NovoEight	0	0	
	Afstyla	314 500	3	104 833.3
	SHL REC FVIII total	**2 661 500**	**44**	**60 488.6**
	Standard FVIII total	*3 459 250*	*50*	*69 185.0*
	Elocta	4 713 493	52	90 644.1
	Adynovi	2 023 000	8	252 875.0
	Jivi	532 000	2	266 000.0
	Esperoct	259 000	3	86 333.3
	EHL REC FVIII total	*7 463 493*	*64*	*116 617.1*
	FVIII total	**10 922 743**	**108**	**101 136.5**
FIX (IU)	Immunine	0	0	
	Octanine	0	0	
	FIX PD total	**0**	**0**	
	Rixubis	238 500	4	59 625.0
	Benefix	0	0	
	FIX REC total	**238 500**	**4**	**59 625.0**
	Standard FIX total	*238 500*	*4*	*59 625.0*
	Alprolix	900 000	14	64 285.7
	Other–rec EHL FIX	0	0	
	EHL REC FIX total	*900 000*	*14*	*64 285.7*
	FIX total	**1 138 500**	**16**	**71 156.3**
By-pass	NovoSeven (mg)	1 373.0	2	686.5
Emicizumab	Hemlibra s.c. (mg)	72 454	48	1 509.5

Consumption of drugs

Adults

	Drug (IU)	Total annual consumption	Number of treated persons	Average annual consumption per treated person
FVIII (IU)	Fanhdi	1 225 900	26	47 150.0
	Octanate	223 000	5	44 600.0
	Wilate	0	0	
	Immunate	1 000	2	500.0
	Haemate P	0	0	
	Other plasma-derived	300	1	300.0
	PD FVIII total	**1 449 900**	**28**	**51 782.1**
	Advate	5 817 250	61	95 364.8
	NUWIQ	1 679 500	10	167 950.0
	Kovaltry	28 000	1	28 000.0
	NovoEight	986 500	5	197 300.0
	Afstyla	2 000	1	2 000.0
	SHL REC FVIII total	**7 815 750**	**72**	**108 552.1**
	Standard FVIII total	*9 265 650*	*98*	*94 547.4*
	Elocta	9 828 081	47	209 108.1
	Adynovi	9 392 700	39	240 838.5
	Jivi	2 795 000	12	232 916.7
	Esperoct	1 872 200	7	267 457.1
	EHL REC FVIII total	*23 675 981*	*99*	*239 151.3*
	FVIII total	**32 941 631**	**187**	**176 158.5**
FIX (IU)	Immunine	304 200	11	27 654.5
	Octanine	299 500	4	74 875.0
	FIX PD total	**603 700**	**14**	**43 121.4**
	Rixubis	351 000	2	175 500.0
	Benefix	10 000	1	10 000.0
	FIX REC total	**361 000**	**3**	**120 333.3**
	Standard FIX total	*964 700*	*17*	*56 747.1*
	Alprolix	2 772 870	26	106 648.8
	Other–rec EHL FIX	471 600	8	58 950.0
	EHL REC FIX total	*3 244 470*	*29*	*111 878.3*
	FIX total	**4 209 170**	**40**	**105 229.3**
By-pass	NovoSeven (mg)	5.0	1	5.0
Emicizumab	Hemlibra s.c. (mg)	92 552	14	6 610.8