The current status of care for persons with haemophilia in The Czech Republic based on representative sample data of Czech haemophilia population in 2011

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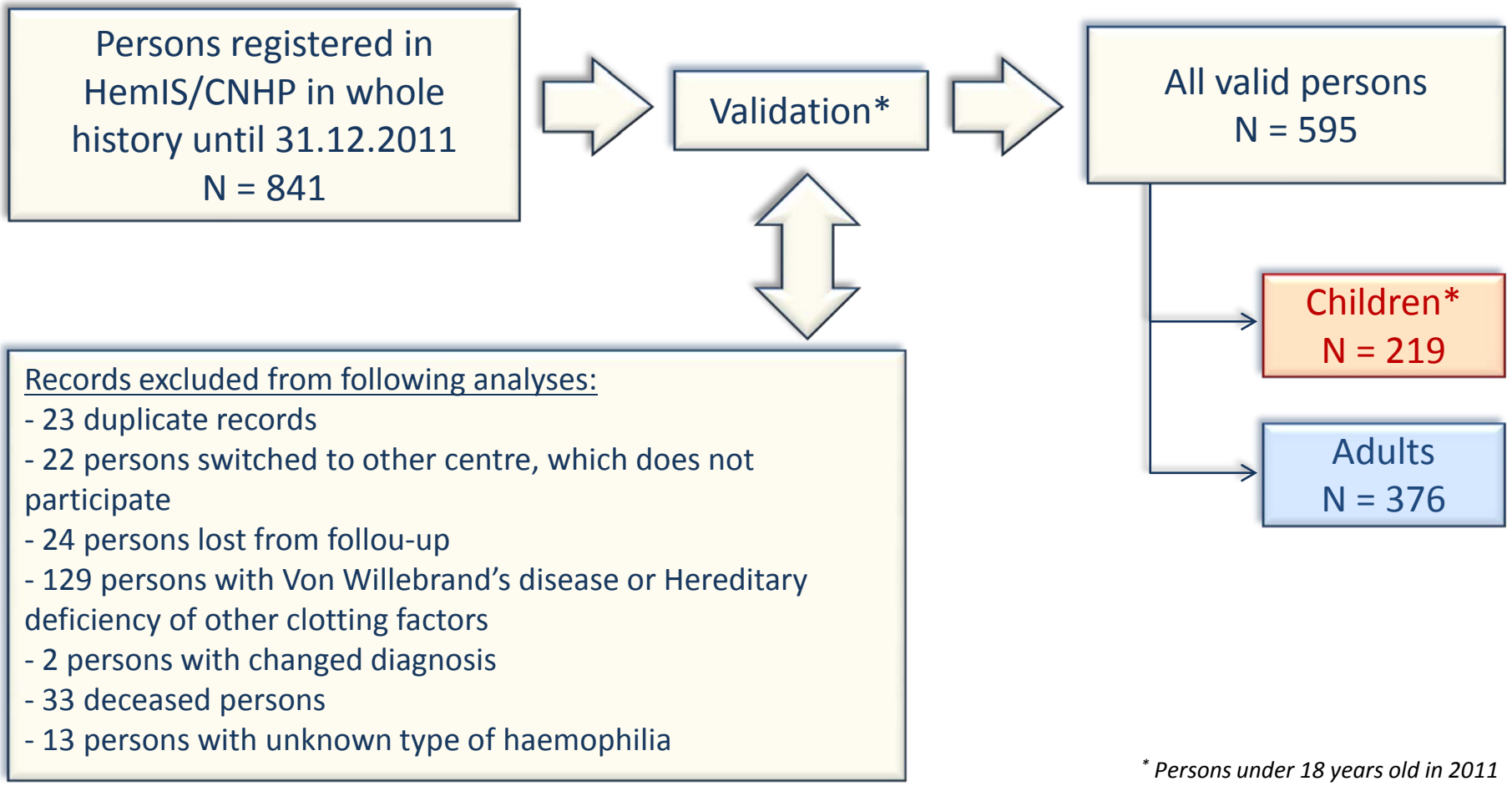
for

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

Sample size, valid records



* *Persons under 18 years old in 2011*

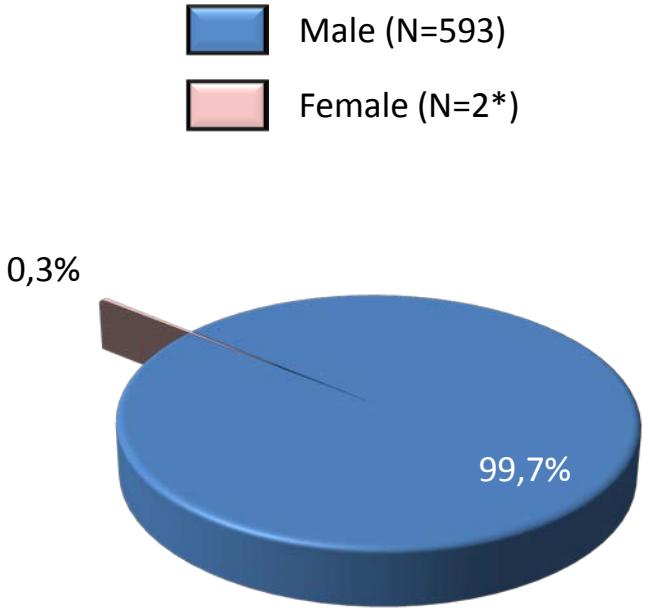
Participating centres in CNHP

Paediatric centres	Valid persons N	%
FN Motol – Dpt. of Pediatric Haematology and Oncology	87	14.6
FN Brno – CUH – Dpt. of Pediatric Haematology	37	6.2
FNHK – Dpt. of Pediatric Medicine	35	5.9
FN Ostrava – Dpt. of Pediatric Medicine	29	4.9
UnL – Pediatric Dpt. – Haematology	24	4.0
FN Olomouc – Dpt. of Pediatric Medicine	13	2.2
FN Plzeň – Pediatric Dpt.	10	1.7
CB – Pediatric Dpt.	7	1.2

Adult centres	Valid persons N	%
FN Brno – Dpt .of Clin Haematology	122	20.5
FN Ostrava – Blood centre	70	11.8
FN Olomouc – Haemato-Oncology Dpt.	54	9.1
FN Plzen – UKBH	34	5.7
CB – Dpt .of Clin Haematology	27	4.5
KN Liberec – Dpt .of Clin Haematology	21	3.5
FNHK – Dpt .of Clin Haematology	9	1.5
UnL – Dpt .of Clin Haematology	8	1.3
Plzen – Health Centre – Haematology	7	1.2
Kolin – Haematology and Transfusion Dpt.	1	0.2

All
N=595

Sex

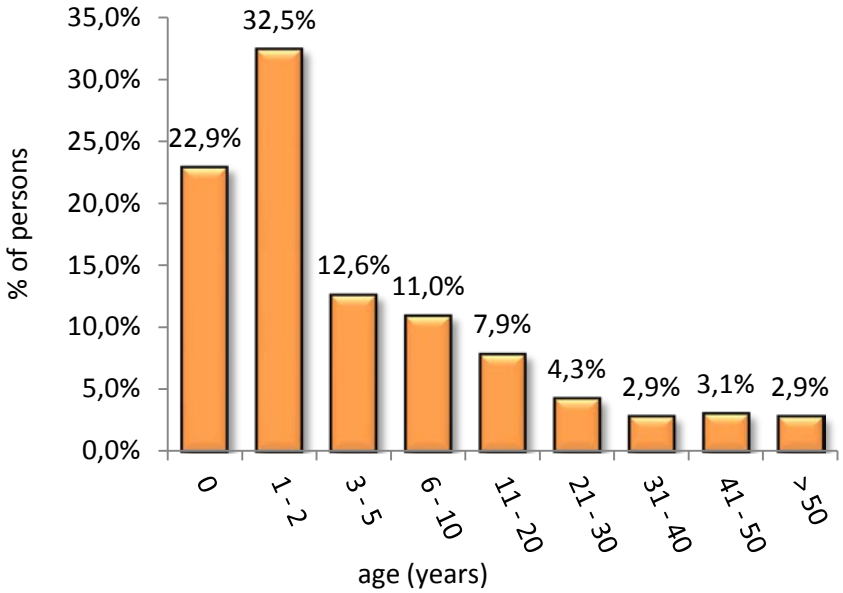


* *both adult*

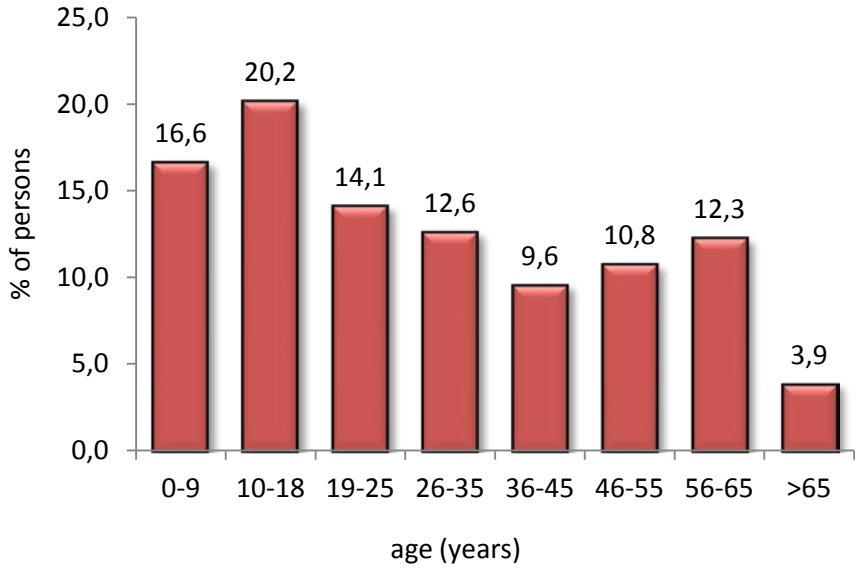
All
N=595

Age

	Age at diagnosis (years)
N	*419**
Mean	*8.1*
Median (min - max)	*2 (0 – 73)*



	Current age (years)
N	*595*
Mean	*30.0*
Median (min - max)	*25 (0 – 89)*



** Missing information on year of diagnosis in 176 persons.*

Persons with haemophilia with inhibitor

All
N=595

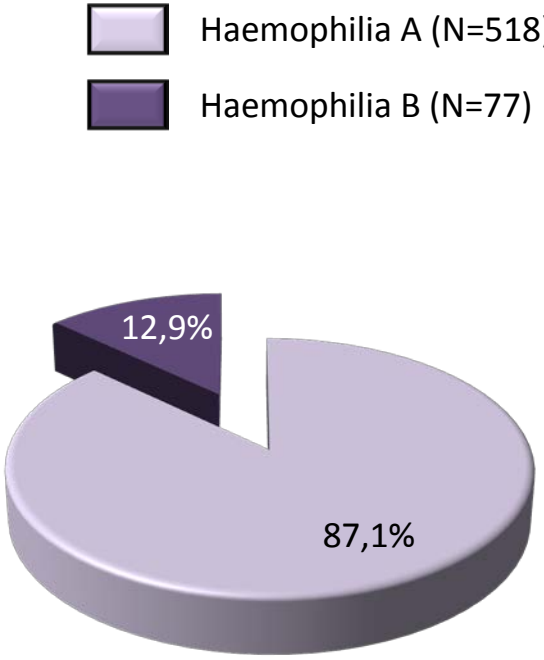
- inhibitor was recorded in 11 persons in year 2011
- other 3 persons have recorded inhibitor in 2010 (data from 2011 are not available)

currently recorded 14 persons with inhibitor (7 children and 7 adults)

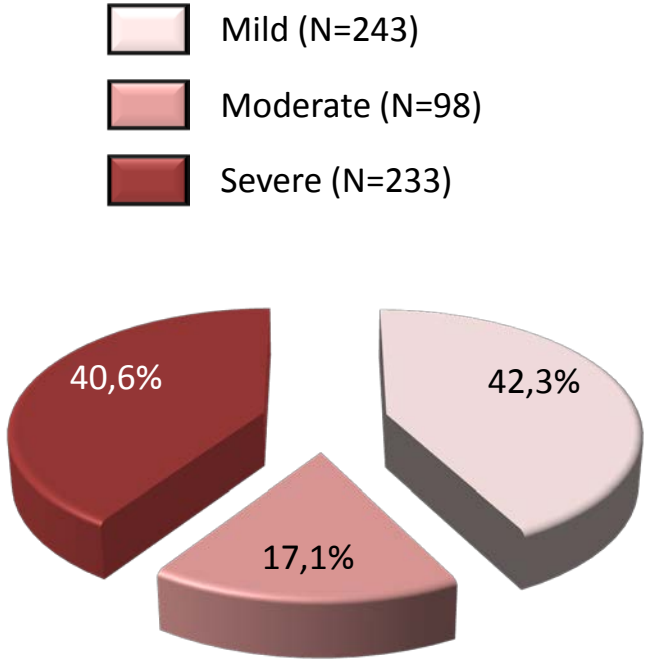
All
N=595

Type and severity of haemophilia I

Type of haemophilia



Severity of haemophilia (N=574*)

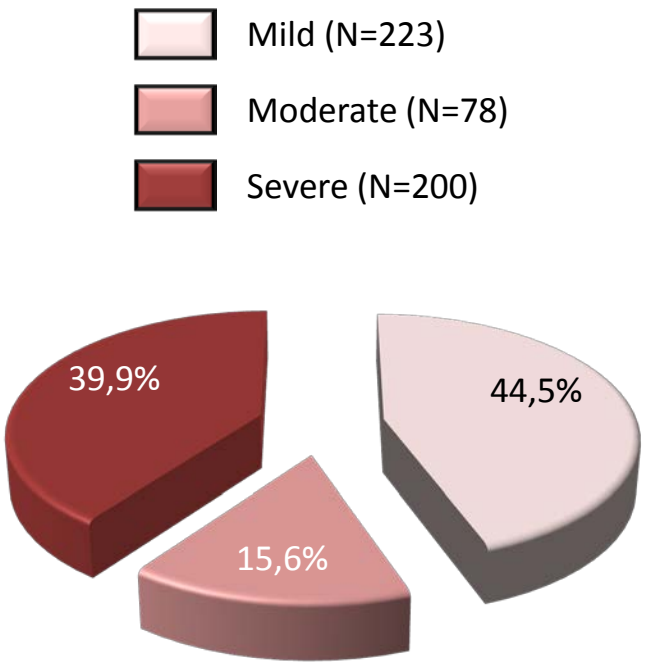


* *Severity of haemophilia not known in 21 persons.*

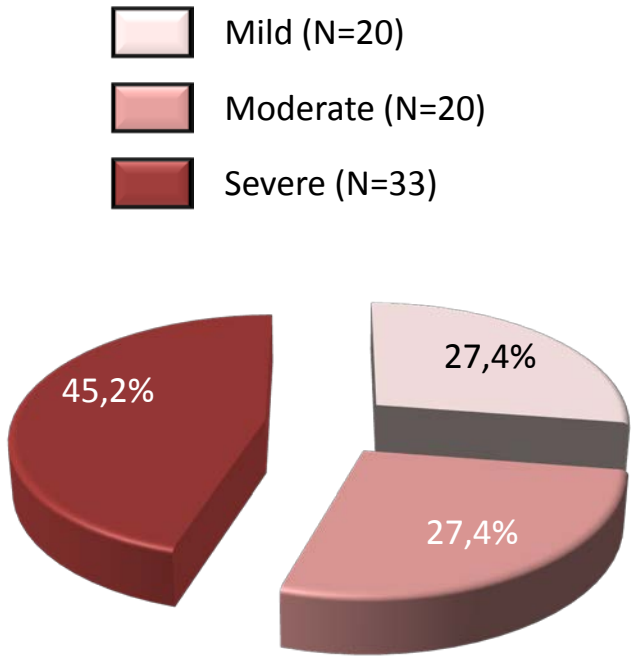
All
N=595

Type and severity of haemophilia II

Haemophilia A (N=501[1)])



Haemophilia B (N=73[2)])



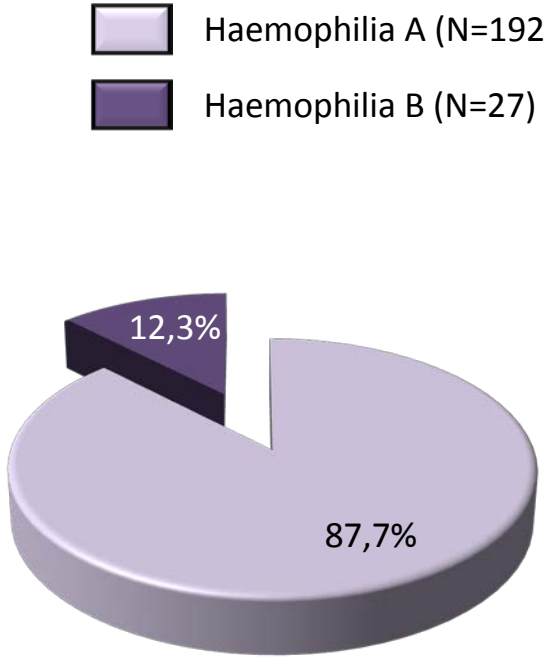
[1)] *Severity not known in 17 persons with haemophilia A.*

[2)] *Severity not known in 4 persons with haemophilia B.*

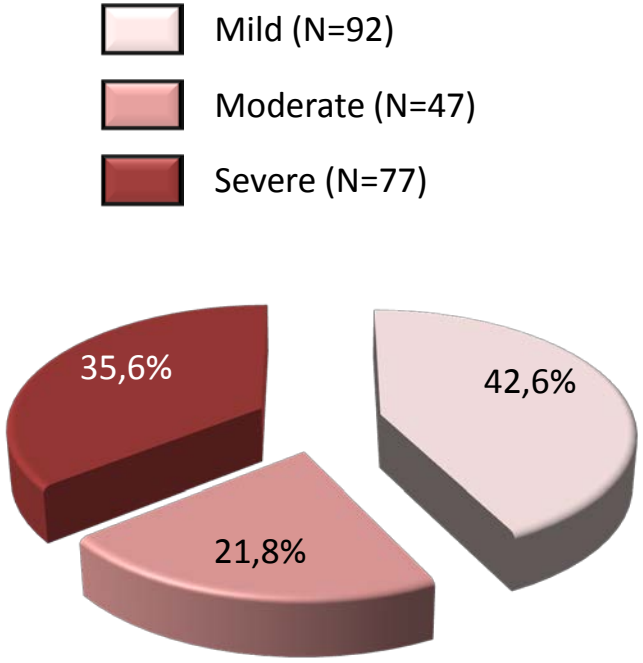
Children
N=219

Type and severity of haemophilia I

Type of haemophilia



Severity of haemophilia (N=216*)

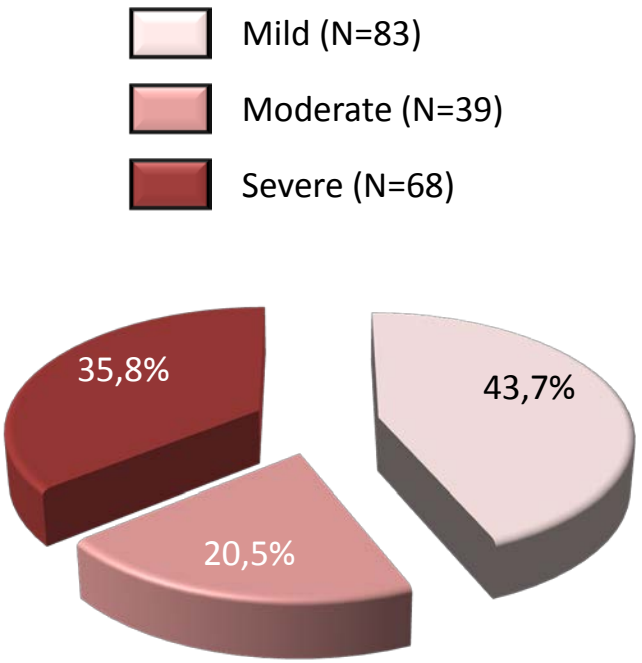


* *Severity of haemophilia not known in 3 children.*

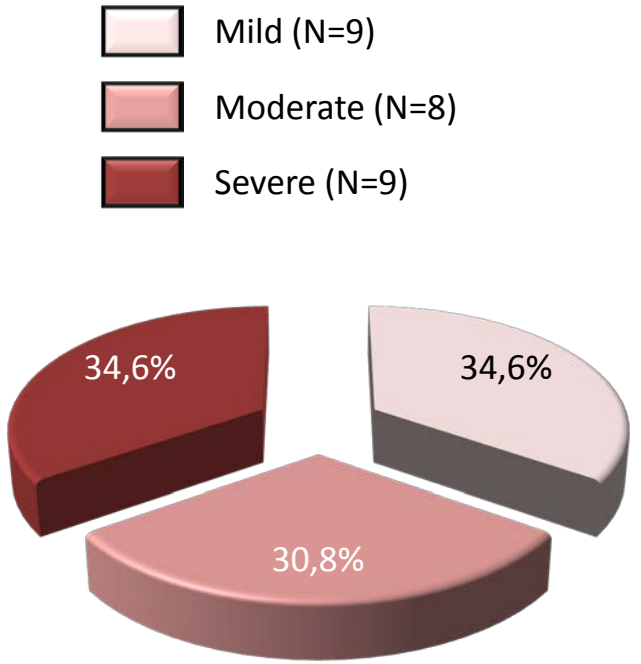
Children
N=219

Type and severity of haemophilia II

Haemophilia A (N=190[1)])



Haemophilia B (N=26[2)])



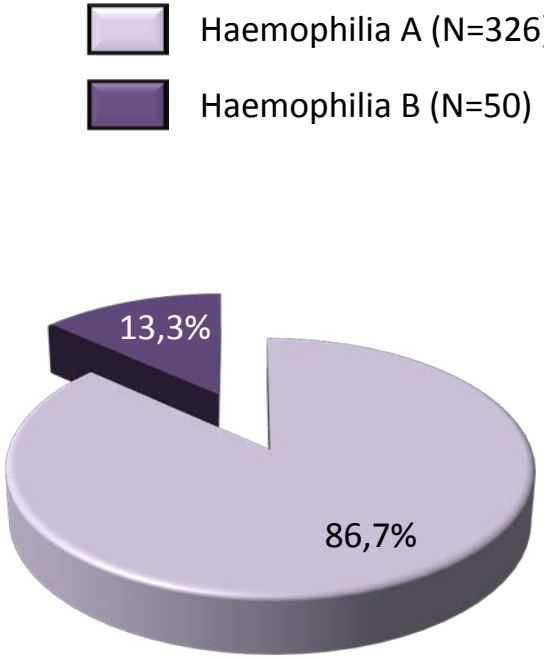
[1)] *Severity not known in 2 children with haemophilia A.*

[2)] *Severity not known in 1 child with haemophilia B.*

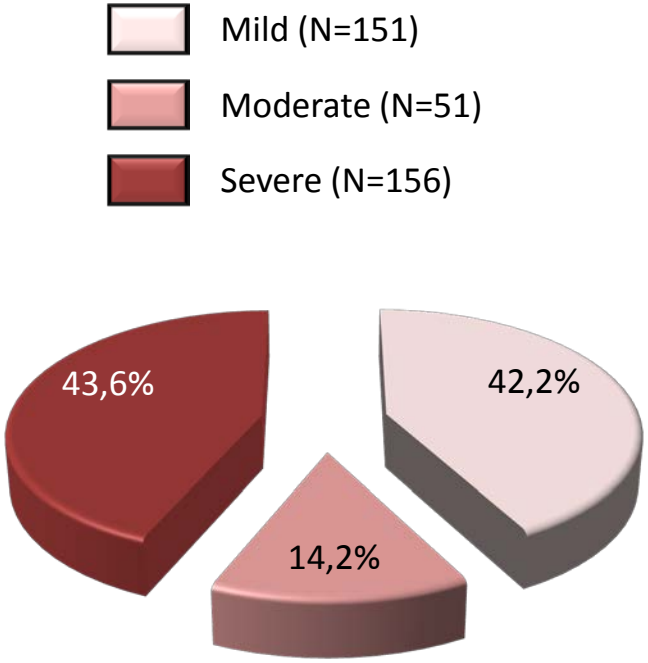
Adults
N=376

Type and severity of haemophilia I

Type of haemophilia



Severity of haemophilia (N=358*)

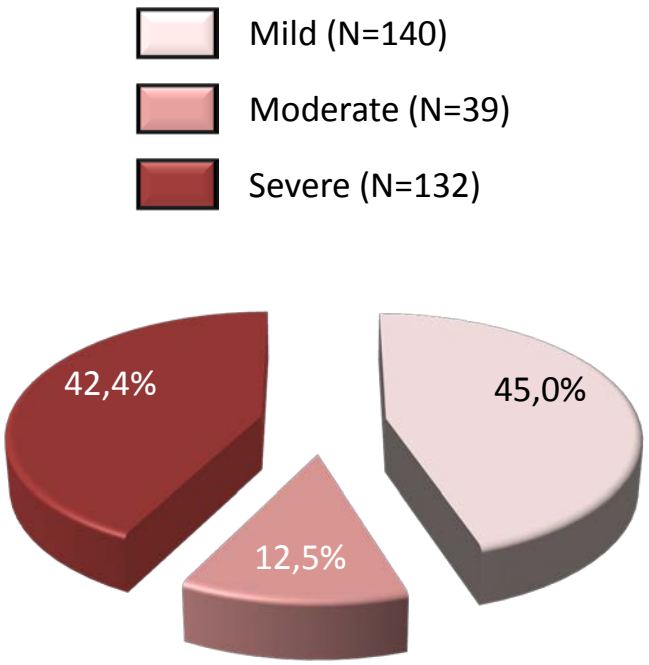


* *Severity of haemophilia not known in 18 adults.*

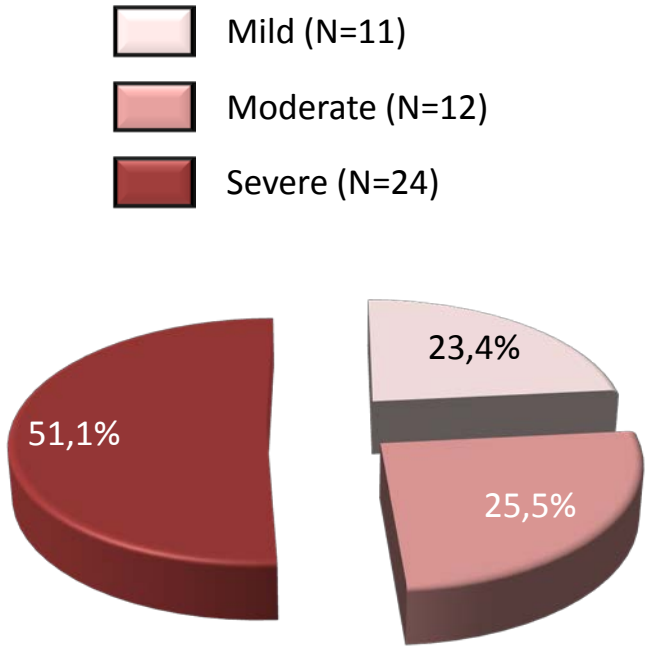
Adults
N=376

Type and severity of haemophilia II

Haemophilia A (N=311[1)])



Haemophilia B (N=47[2)])

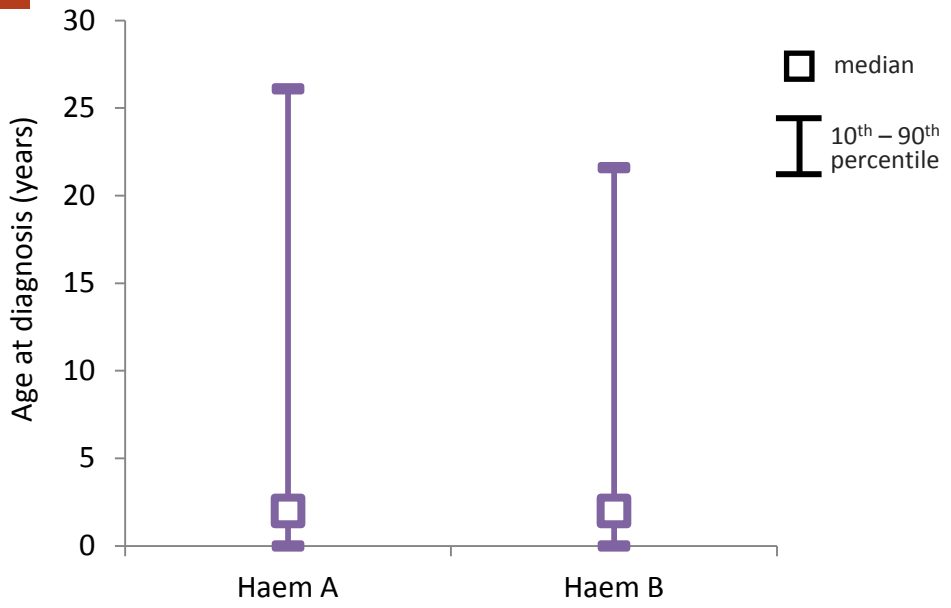


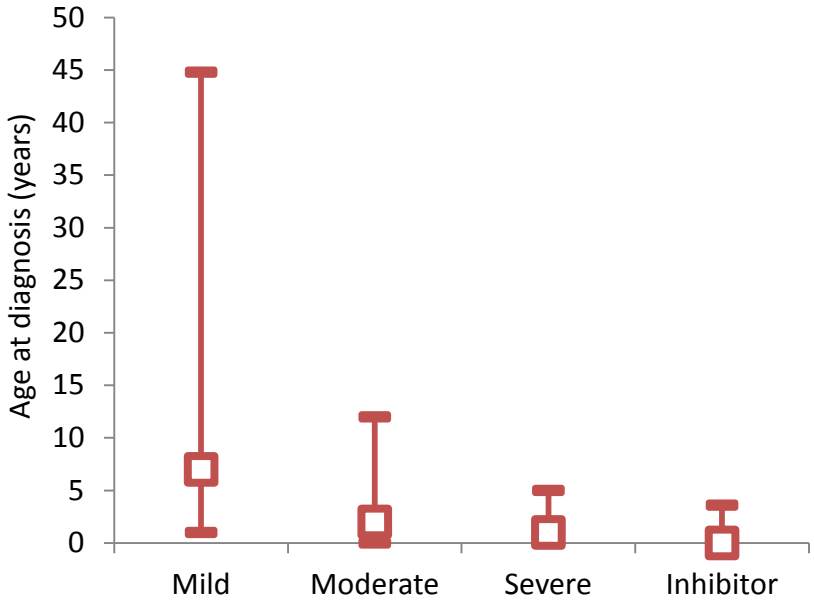
[1)] *Severity not known in 15 adults with haemophilia A.*

[2)] *Severity not known in 3 adults with haemophilia B.*

Age at diagnosis according to type and severity of haemophilia

All
N=595



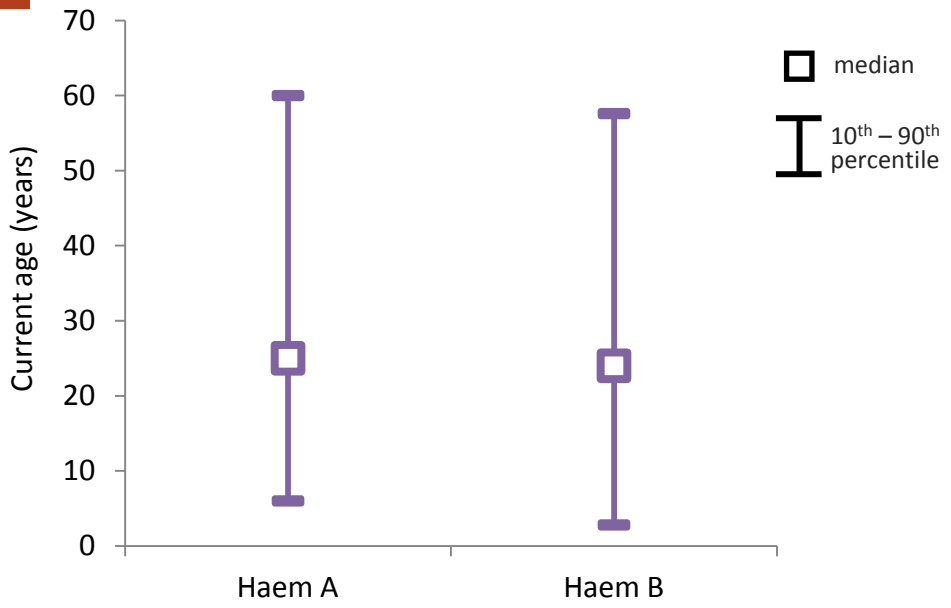


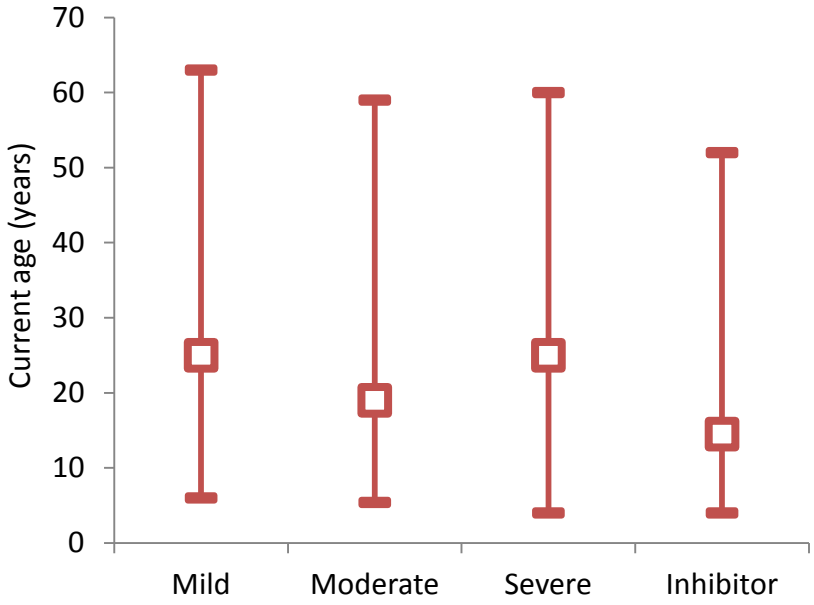
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
361	*58*	**N**	*180*	*76*	*159*	*11*
8.1	*8.2*	**Mean**	*14.4*	*4.4*	*2.1*	*0.9*
2 (0 – 73)	*2 (0 – 3)*	**Median (min – max)**	*7 (0 – 73)*	*2 (0 – 61)*	*1 (0 – 48)*	*0 (0 – 4)*

** including persons with inhibitor*

Current age according to type and severity of haemophilia

All
N=595





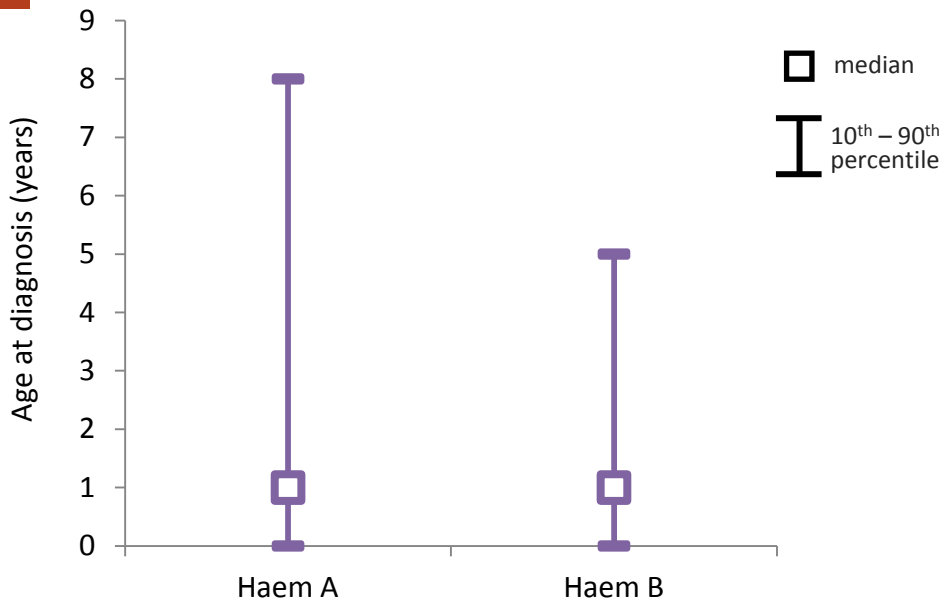
Haemophilia A	Haemophilia B	Current age* (years)	Mild*	Moderate*	Severe*	Inhibitor
518	*77*	**N**	*243*	*98*	*233*	*14*
30.2	*28.8*	**Mean**	*31.1*	*27.0*	*29.4*	*18.9*
25 (0 – 89)	*24 (1 – 68)*	**Median (min – max)**	*25 (0 – 89)*	*19 (1 – 65)*	*25 (0 – 72)*	*14.5 (4 – 55)*

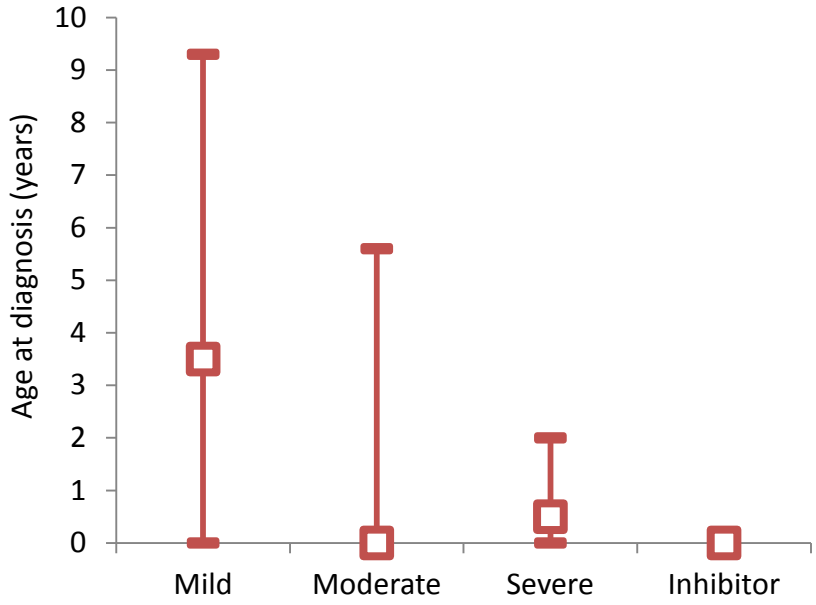
**Current age = age reached in year 2011*

** including persons with inhibitor*

Age at diagnosis according to type and severity of haemophilia

Children
N=219



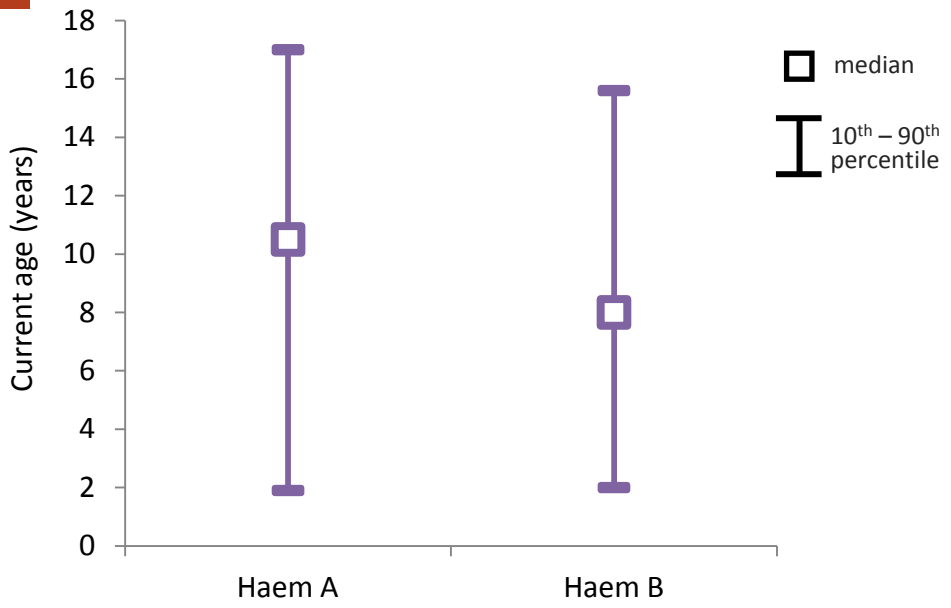


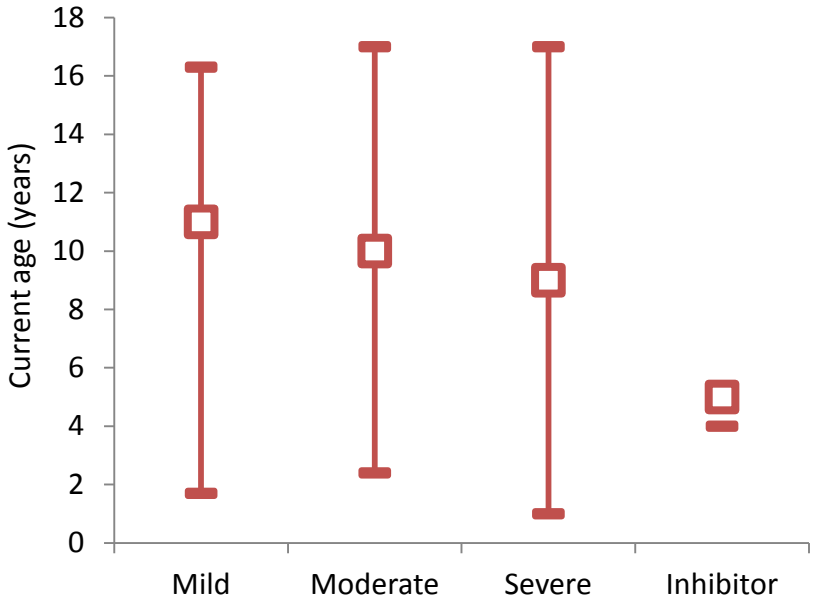
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
138	*23*	**N**	*66*	*33*	*62*	*6*
2.5	*1.8*	**Mean**	*4.1*	*1.9*	*0.9*	*1*
1 (0 – 16)	*1 (0 ––13)*	**Median (min – max)**	*3.5 (0 – 13)*	*0 (0 – 16)*	*0.5 (0 – 11)*	*0 (0 – 4)*

** including persons with inhibitor*

Current age according to type and severity of haemophilia

Children
N=219





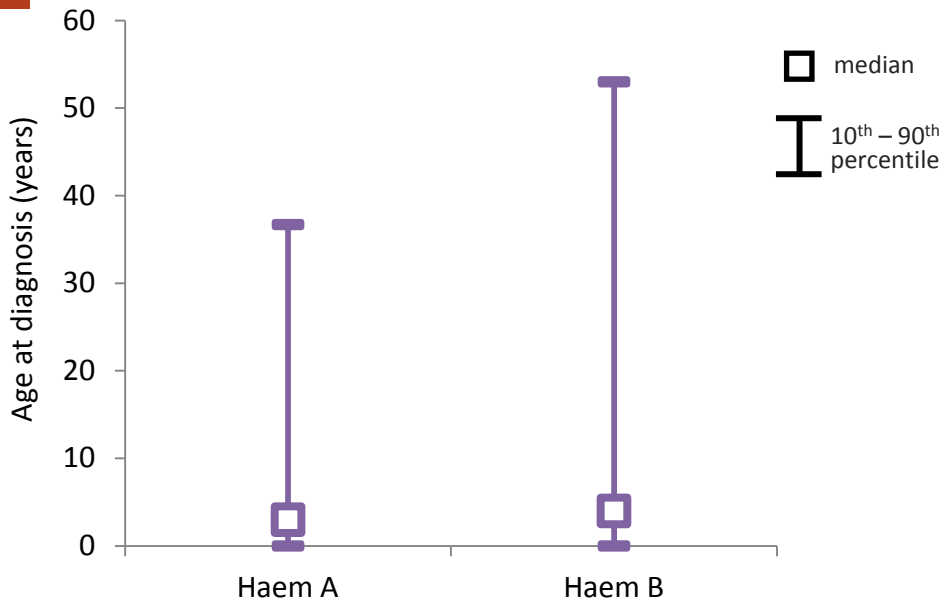
Haemophilia A	Haemophilia B	Current age* (years)	Mild*	Moderate*	Severe*	Inhibitor
192	*27*	**N**	*92*	*47*	*77*	*7*
10.0	*8.7*	**Mean**	*10.0*	*10.5*	*9.4*	*6.0*
10.5 (0 – 18)	*8 (1 – 17)*	**Median (min – max)**	*11 (0 – 18)*	*10 (1 – 18)*	*9 (0 – 18)*	*5 (4 – 10)*

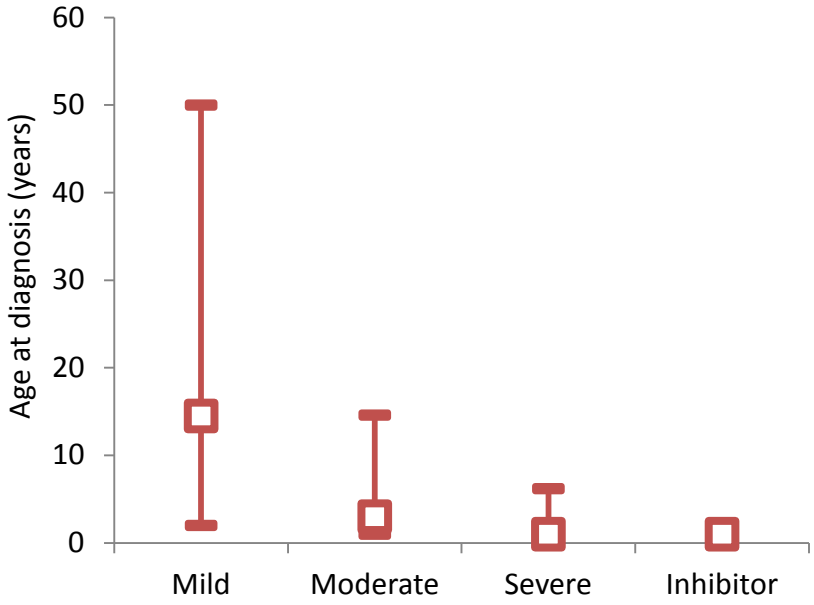
**Current age = age reached in year 2011*

** including persons with inhibitor*

Age at diagnosis according to type and severity of haemophilia

Adults
N=376



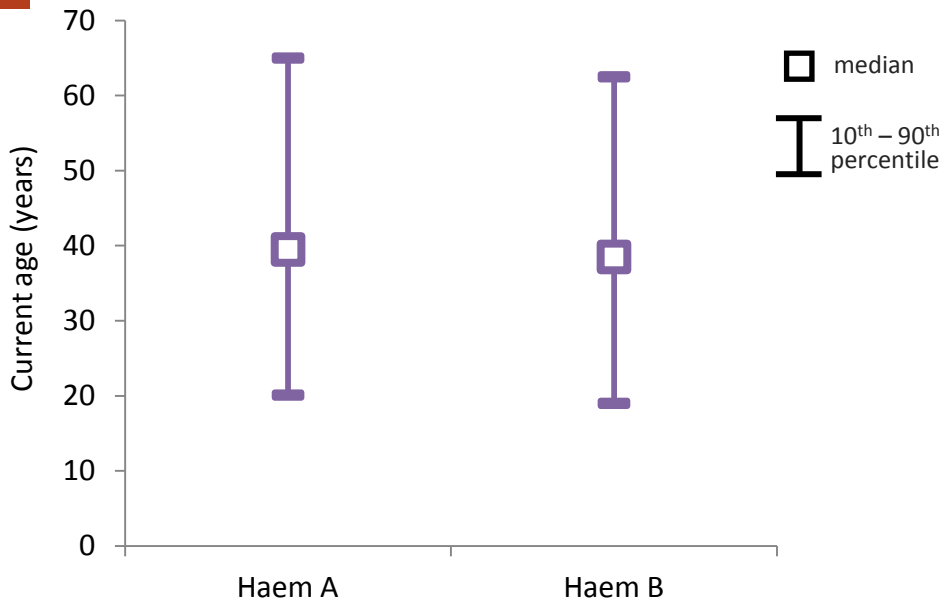


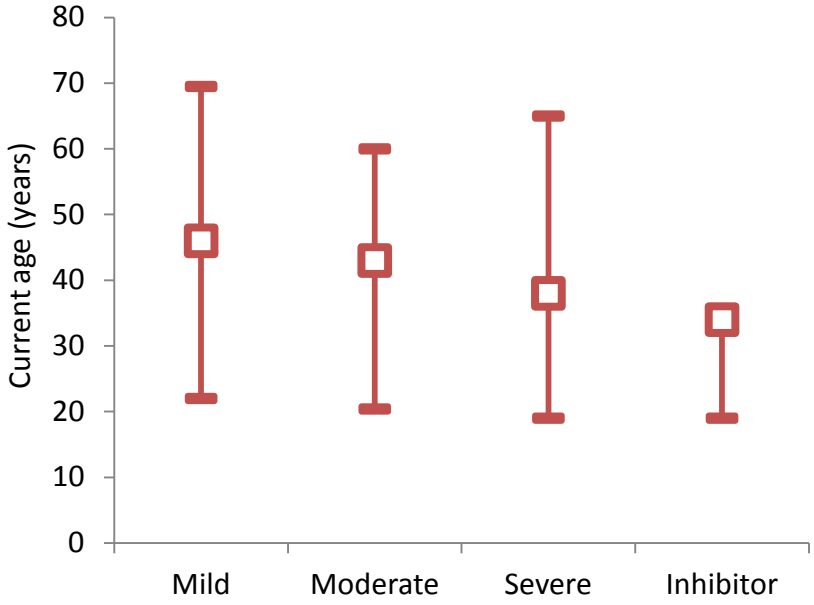
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
223	*35*	**N**	*114*	*43*	*97*	*5*
11.6	*12.3*	**Mean**	*20.4*	*6.3*	*2.9*	*0.8*
3 (0 – 73)	*4 (0 – 63)*	**Median (min – max)**	*14.5 (1 – 73)*	*3 (0 – 61)*	*1 (0 – 48)*	*1 (0 – 2)*

** including persons with inhibitor*

Current age according to type and severity of haemophilia

Adults
N=376





Haemophilia A	Haemophilia B	Current age* (years)	Mild*	Moderate*	Severe*	Inhibitor
326	*50*	**N**	*151*	*51*	*156*	*7*
42.0	*39.6*	**Mean**	*44.0*	*42.2*	*39.3*	*31.7*
39.5 (19 – 89)	*38.5 (19 – 68)*	**Median (min – max)**	*46 (19 – 89)*	*43 (19 – 65)*	*38 (19 – 72)*	*34 (19 – 55)*

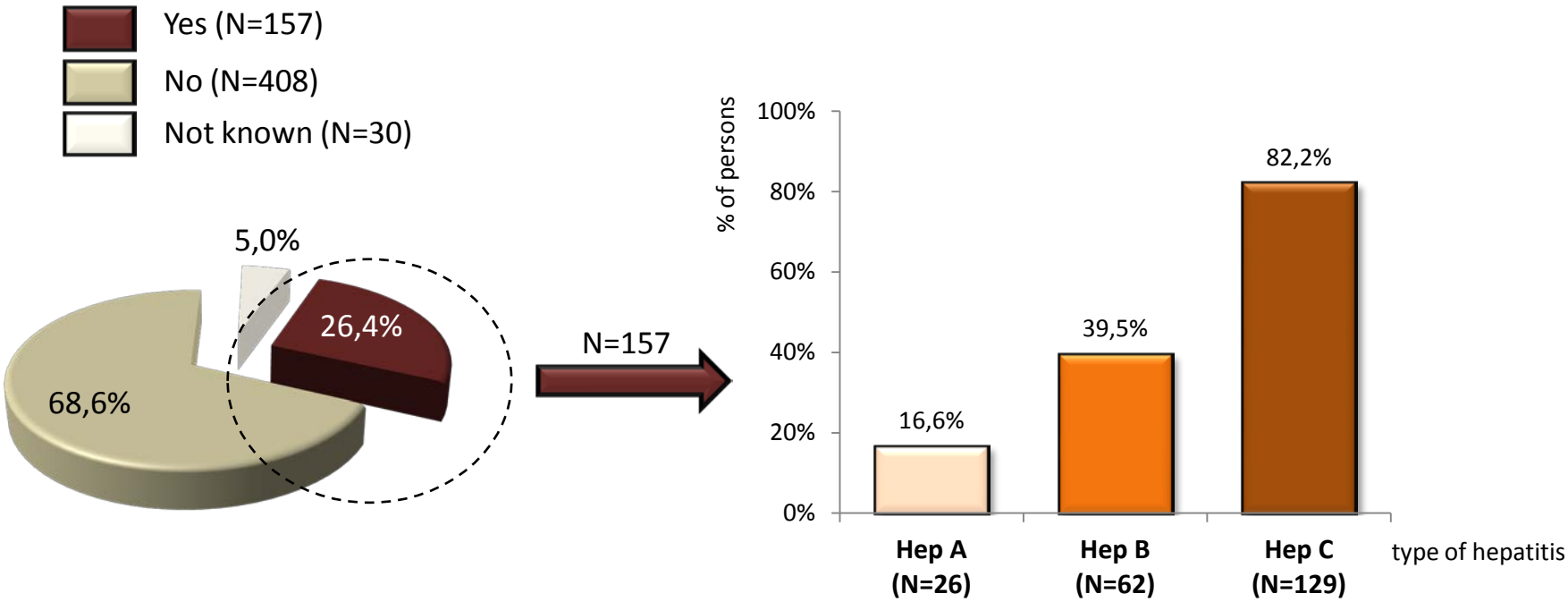
**Current age = age reached in year 2011*

** including persons with inhibitor*

All
N=595

Hepatitis

Experienced hepatitis



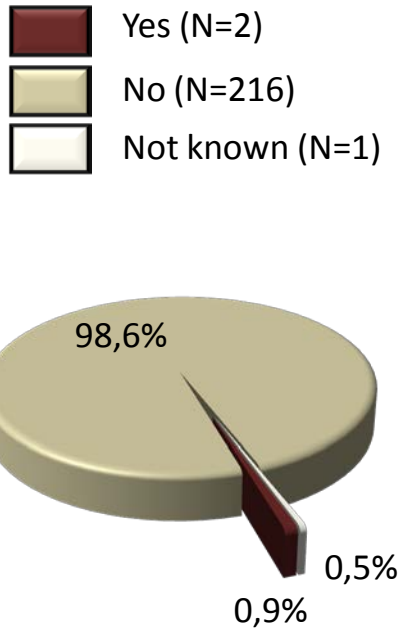
Data from last annual report of each person.

Type of hepatitis not specified in 3 persons. One person may have recorded more types of hepatitis.

Children
N=219

Hepatitis

Experienced hepatitis



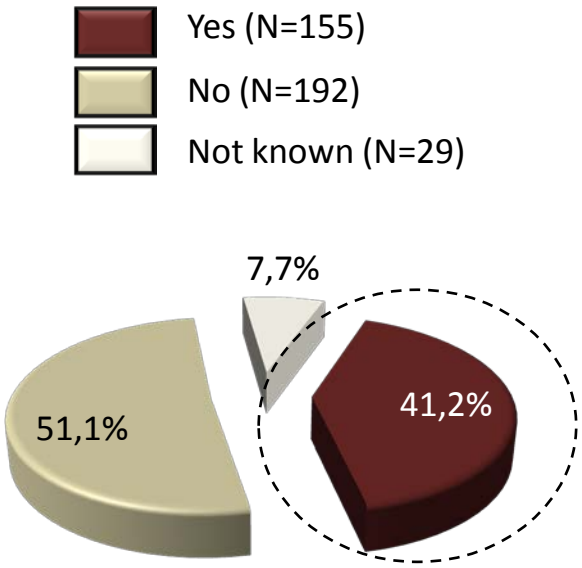
N=2 → *Both children have hepatitis C.*

Data from last annual report of each person.

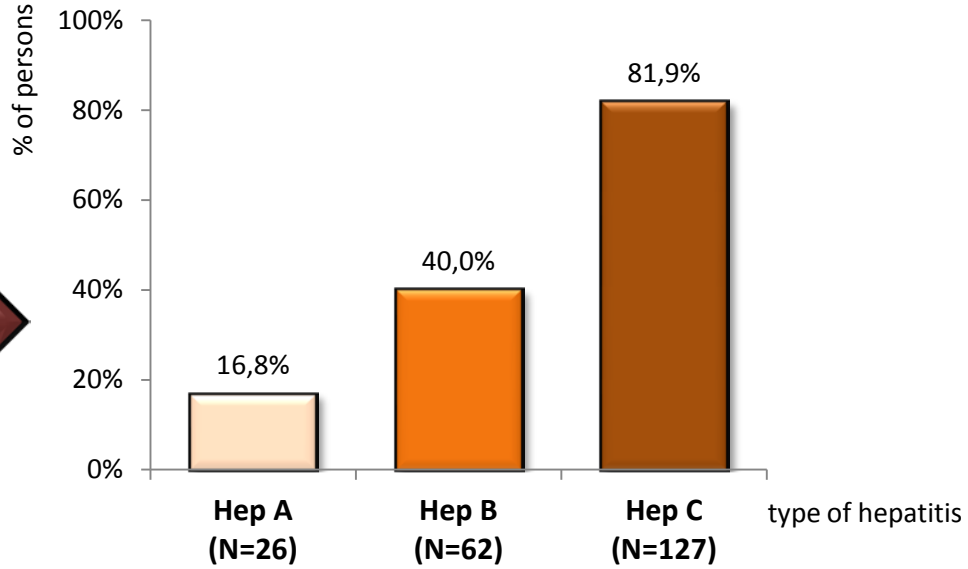
Adults
N=376

Hepatitis

Experienced hepatitis



N=155



Data from last annual report of each person.

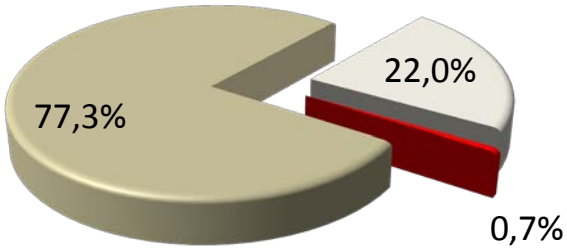
Type of hepatitis not specified in 3 adults. One person may have recorded more types of hepatitis.

All
N=595

HIV

HIV

Positive (N=4)
Negative (N=460)
Not known / not available (N=131)



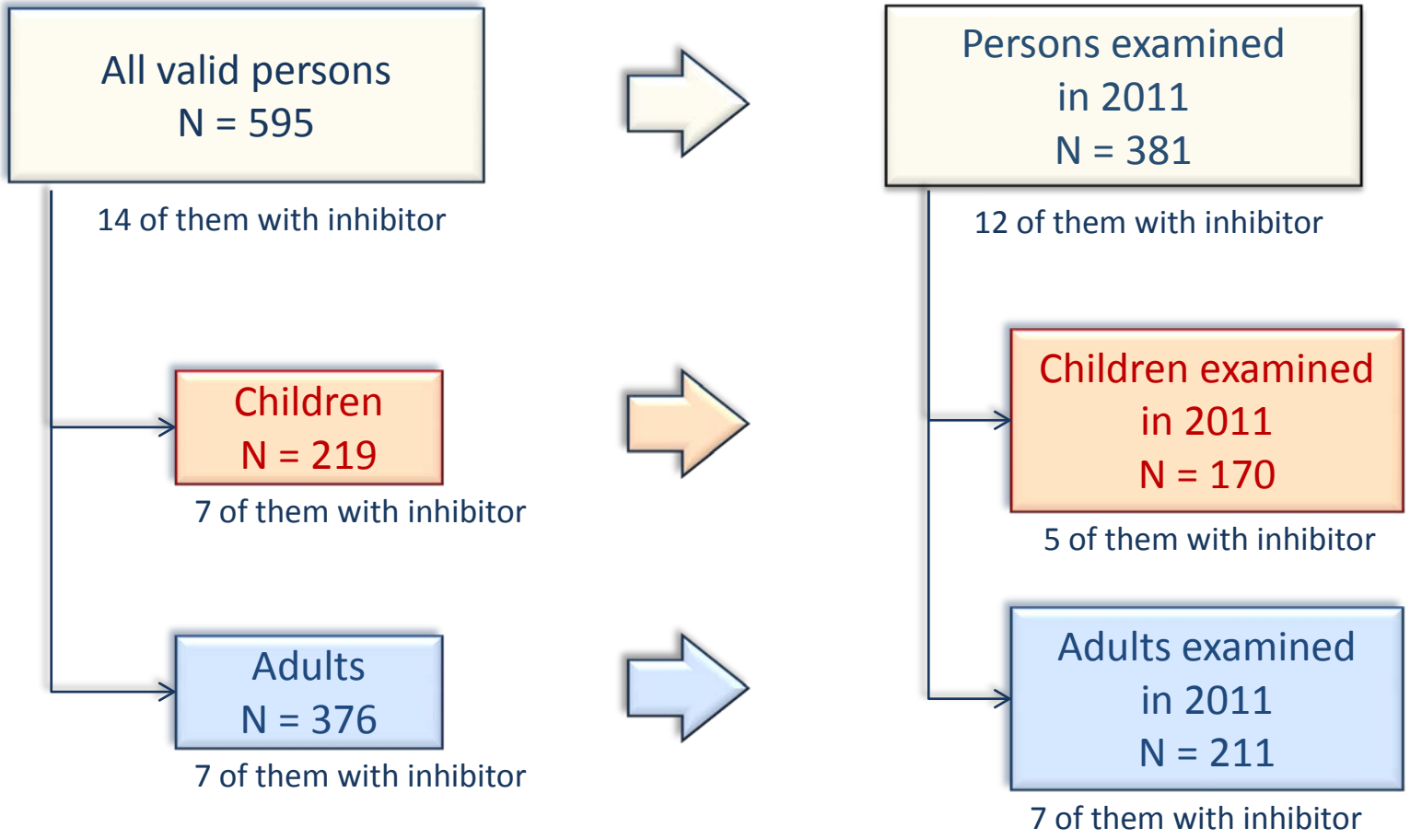
N=4

All 4 HIV-positive persons are adults.

Data retrieved from last annual report of each person.

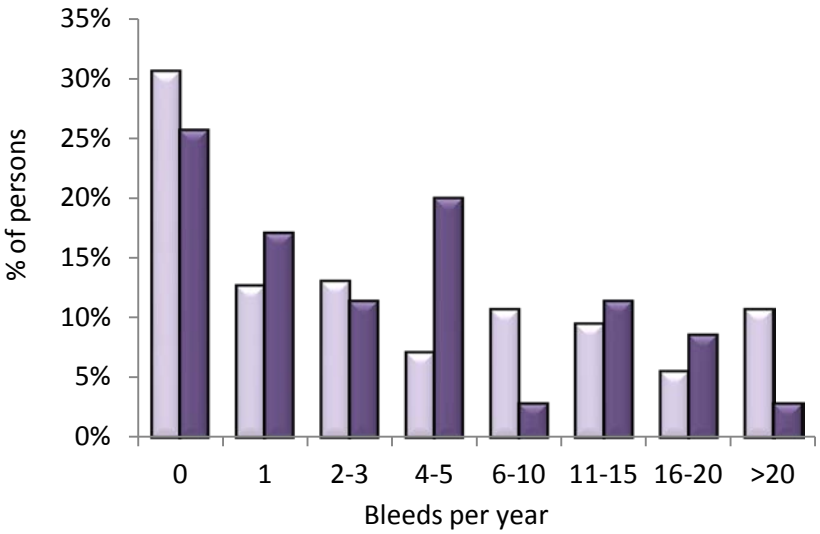
Data of year 2011 – sample size

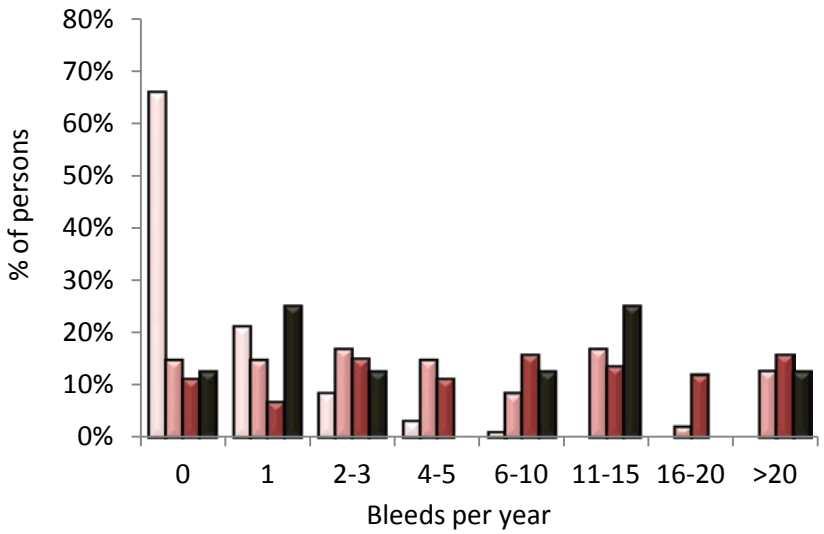
(Data of persons, whose records were updated within 2011)



All
N=381

Bleeding frequency in 2011





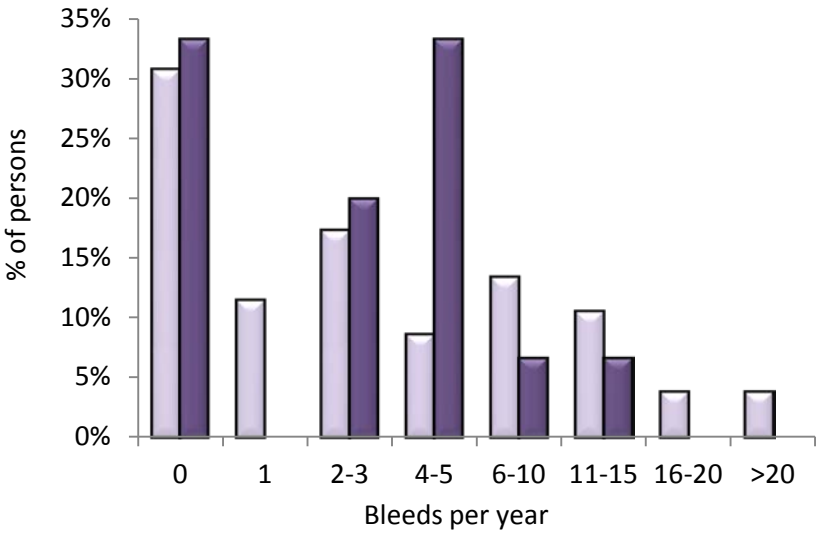
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
252	*35*	**N**	*94*	*48*	*135*	*8*
7.6	*5.6*	**Mean**	*0.7*	*8.9*	*11.5*	*9.4*
2 (0 – 75)	*3 (0 – 25)*	**Median (min – max)**	*0 (0 – 8)*	*4.5 (0 – 41)*	*7 (0 – 75)*	*5 (0 – 36)*

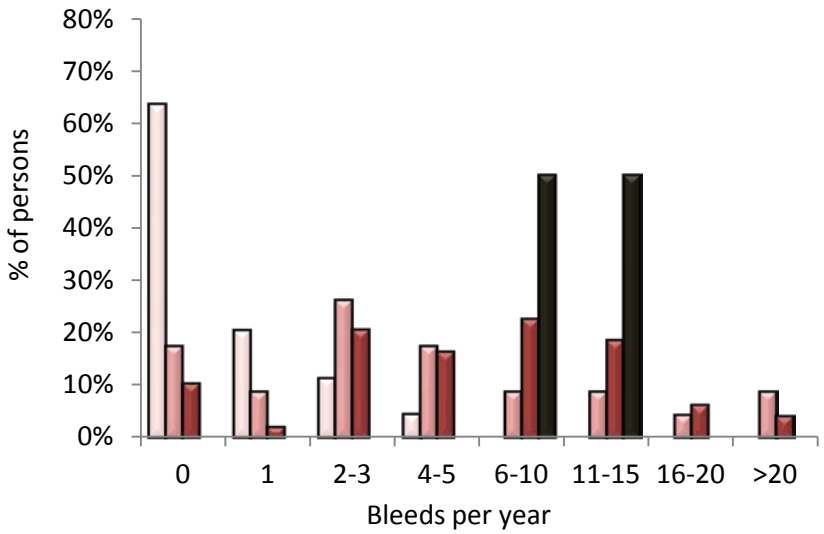
Frequency of bleeding is missing in 94 persons.

** without inhibitor*

Children
N=170

Bleeding frequency in 2011





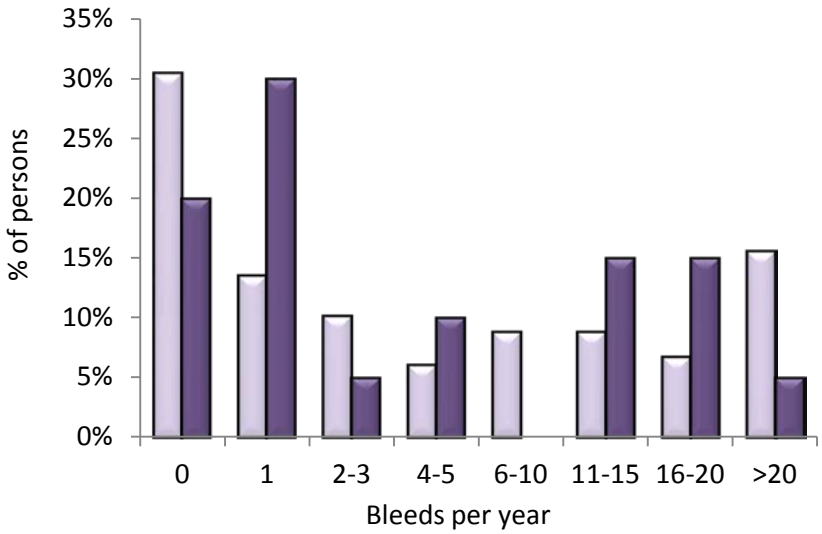
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
104	*15*	**N**	*44*	*23*	*49*	*2*
5.2	*3.4*	**Mean**	*0.7*	*7.2*	*7.6*	*9.5*
2 (0 – 41)	*3 (0 – 14)*	**Median (min – max)**	*0 (0 – 5)*	*3 (0 – 41)*	*6 (0 – 35)*	*9.5 (7 – 12)*

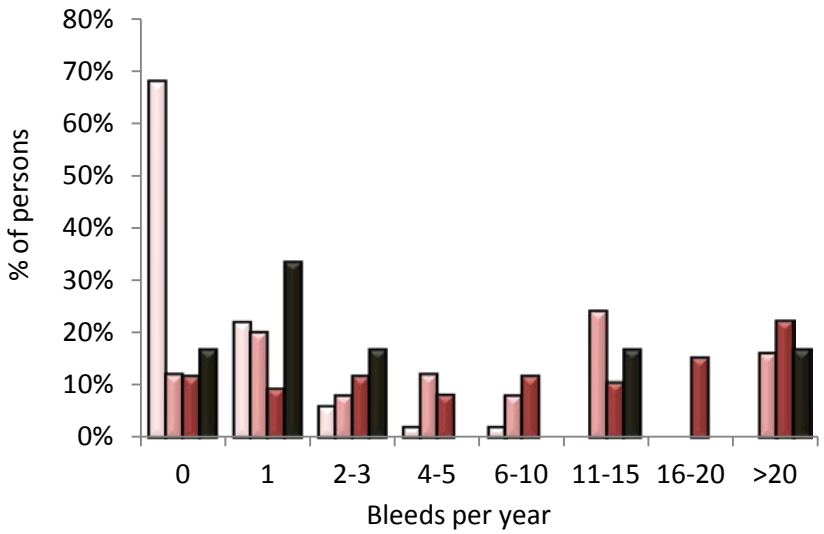
Frequency of bleeding is missing in 51 children.

** without inhibitor*

Adults
N=211

Bleeding frequency in 2011





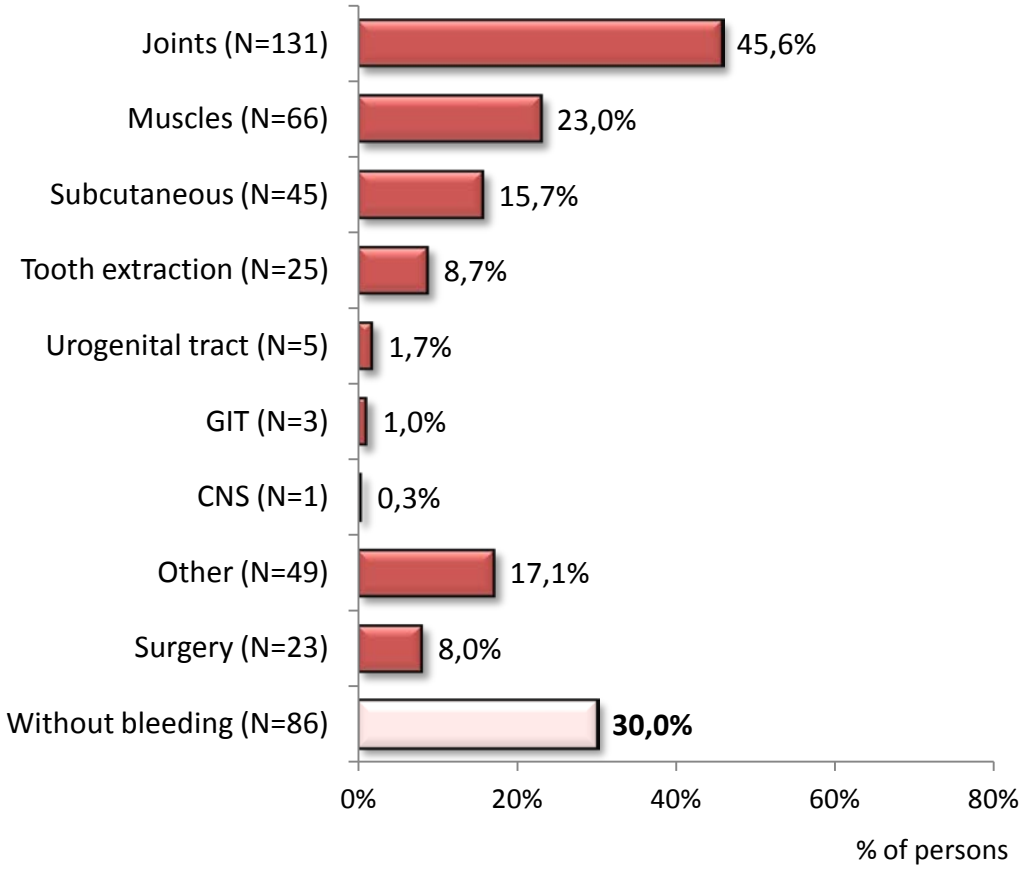
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
148	*20*	**N**	*50*	*25*	*86*	*6*
9.3	*7.3*	**Mean**	*0.6*	*10.5*	*13.7*	*9.3*
3 (0 – 75)	*2 (0 – 25)*	**Median (min – max)**	*0 (0 – 8)*	*5 (0 – 41)*	*10 (0 – 75)*	*2 (0 – 36)*

Frequency of bleeding is missing in 43 adults.

** without inhibitor*

All
N=381

Location of bleeds in 2011

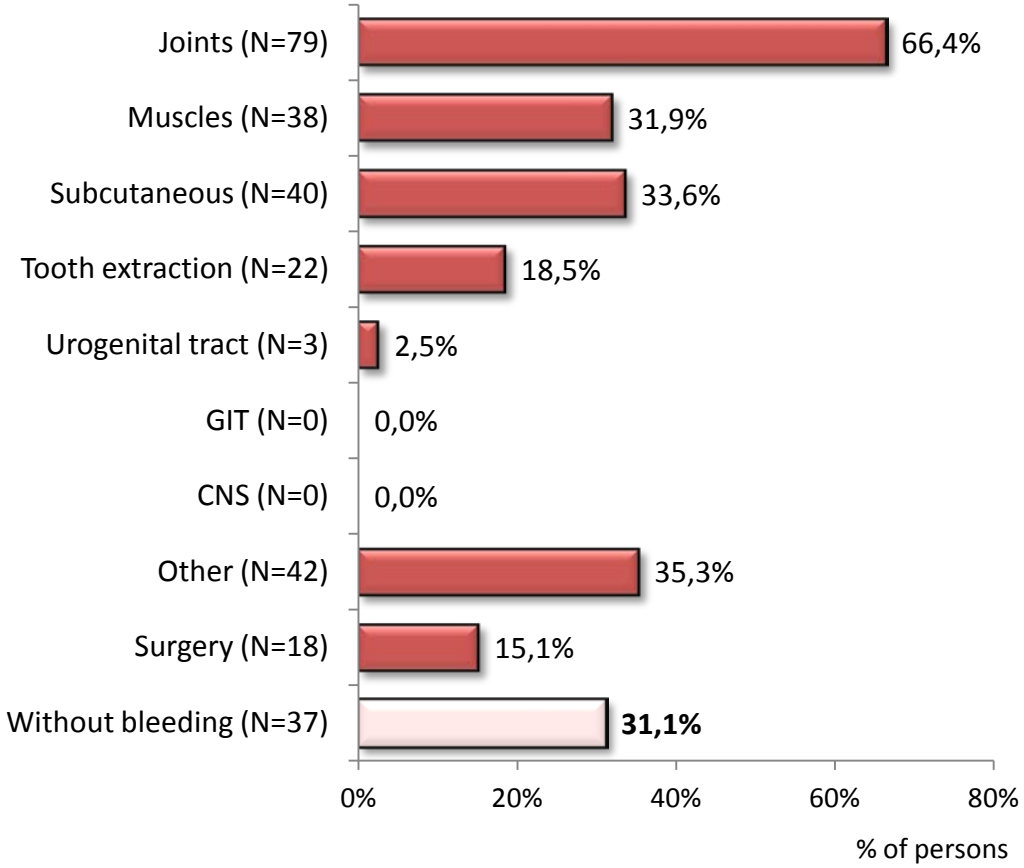


201 (70 %) persons experienced bleeding at least once during the year (2121 bleeds were recorded in total). 86 persons recorded no bleed during year 2011.

Information on frequency of bleeding is missing in 94 persons.

Children
N=170

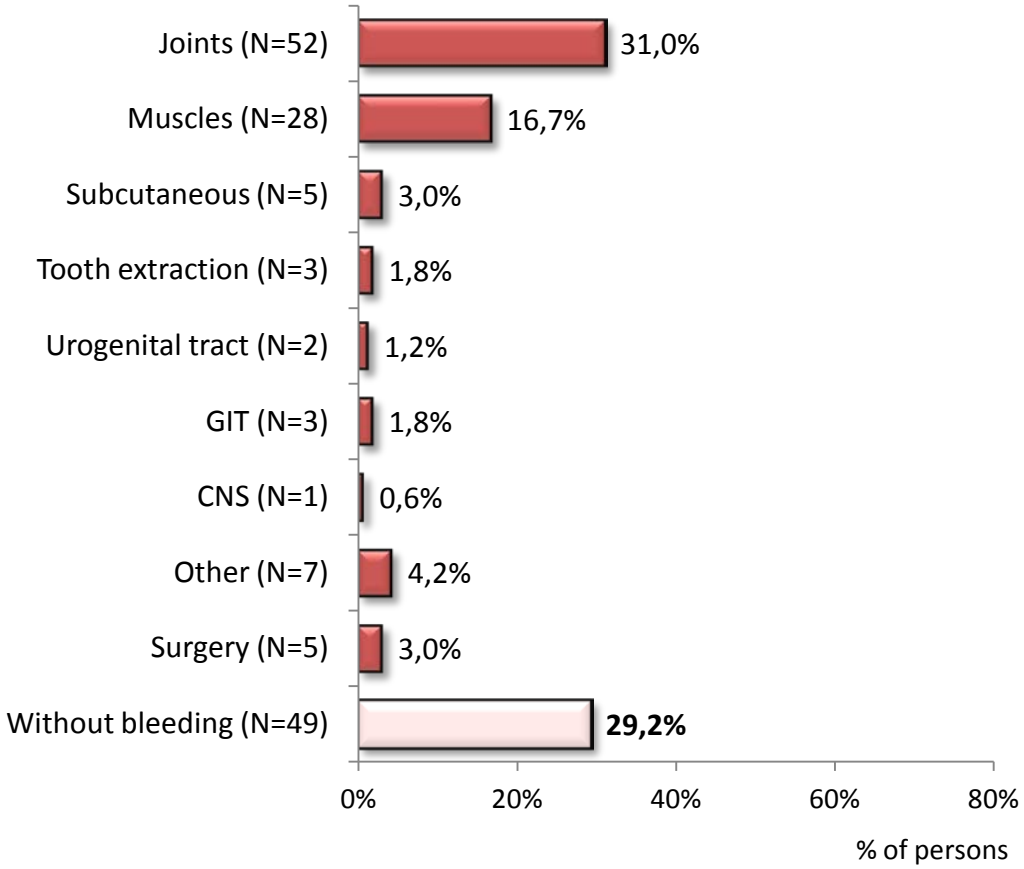
Location of bleeds in 2011



82 (68.9 %) children experienced bleeding at least once during the year (596 bleeds were recorded in total). 37 children recorded no bleed during year 2011.
Information on frequency of bleeding is missing in 51 children.

Adults
N=211

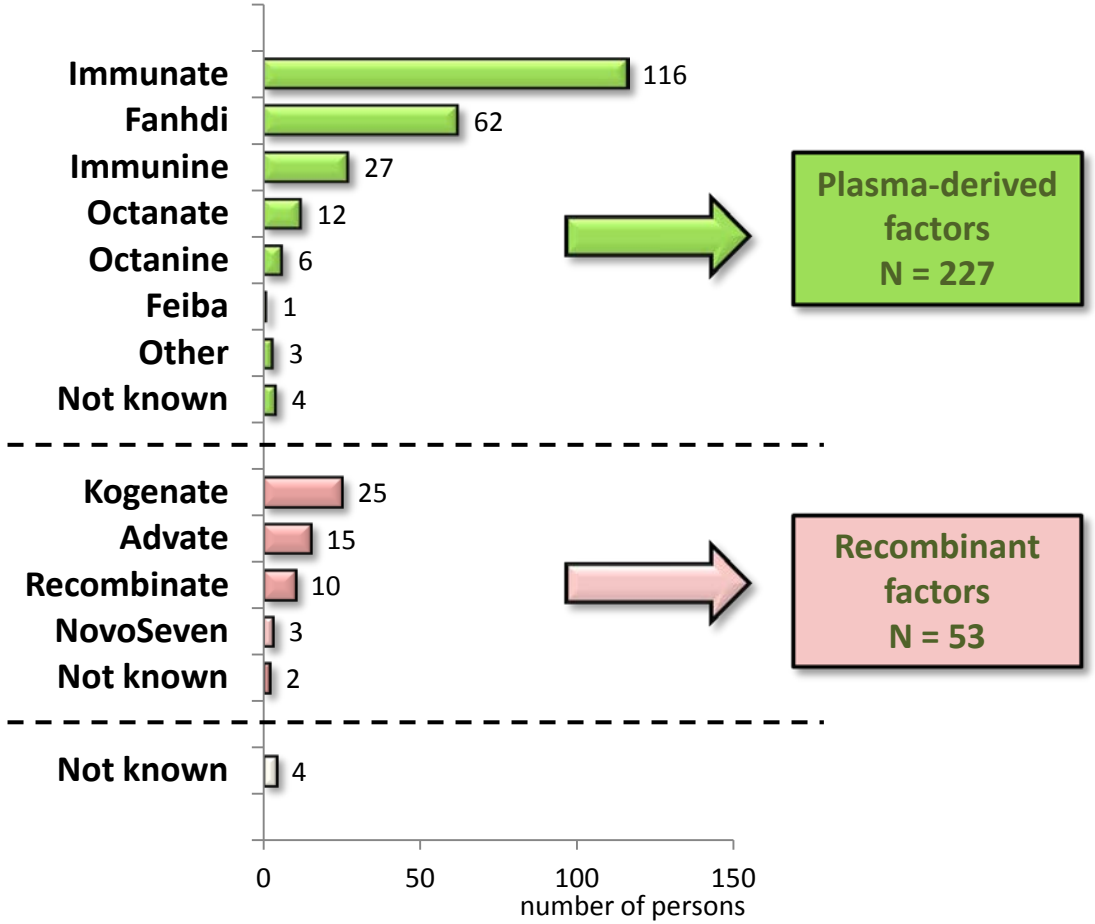
Location of bleeds in 2011



119 (70.8 %) adults experienced bleeding at least once during the year (1525 bleeds were recorded in total). 49 adults have recorded no bleed during year 2011.
Information on frequency of bleeding is missing in 43 adults.

All
N=381

Treatment

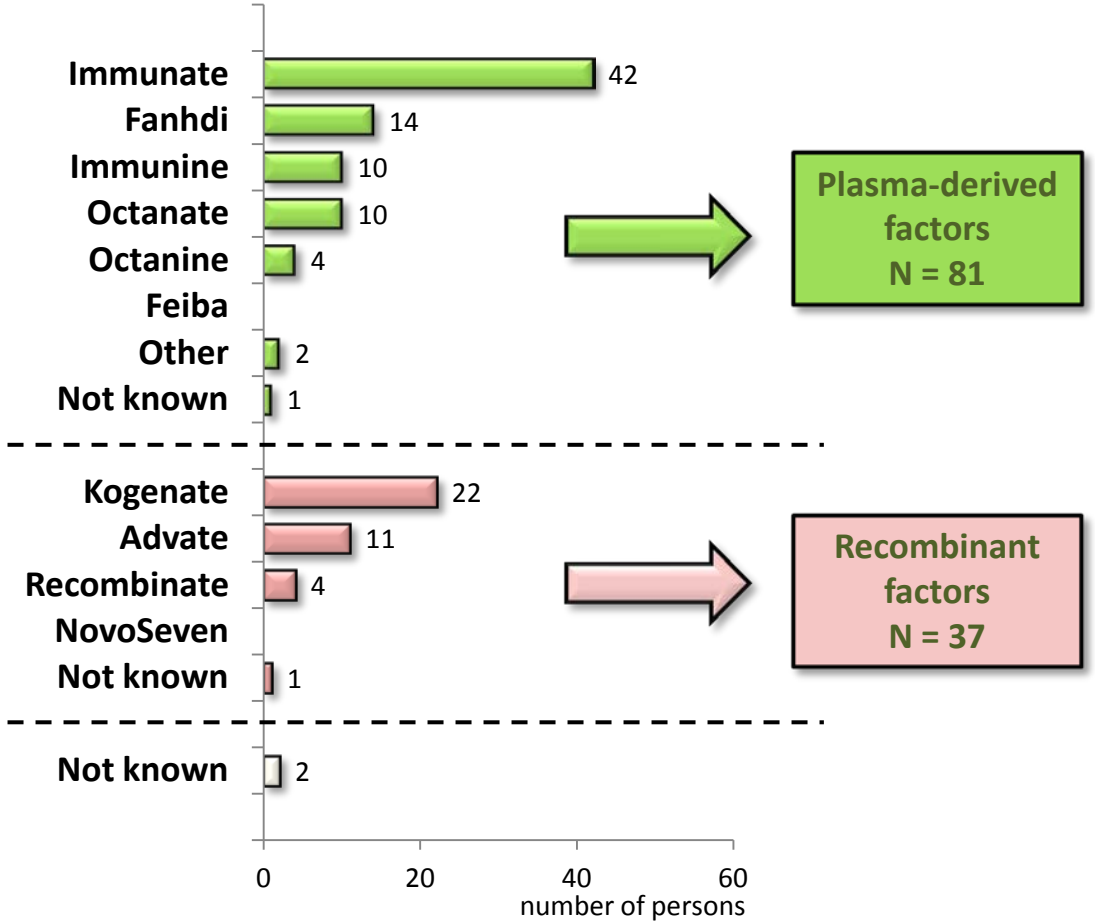


264 (69.3 %) persons were treated in 2011 (33 of them with more different factor concentrates).
Plasma-derived factors were administered more frequently – in 227 persons, recombinant factors in 53 persons and not specified drug in 4 persons.
19 persons were treated with both plasma-derived and recombinant factor.

117 (30.7 %) persons had no treatment in 2011.

Children
N=170

Treatment

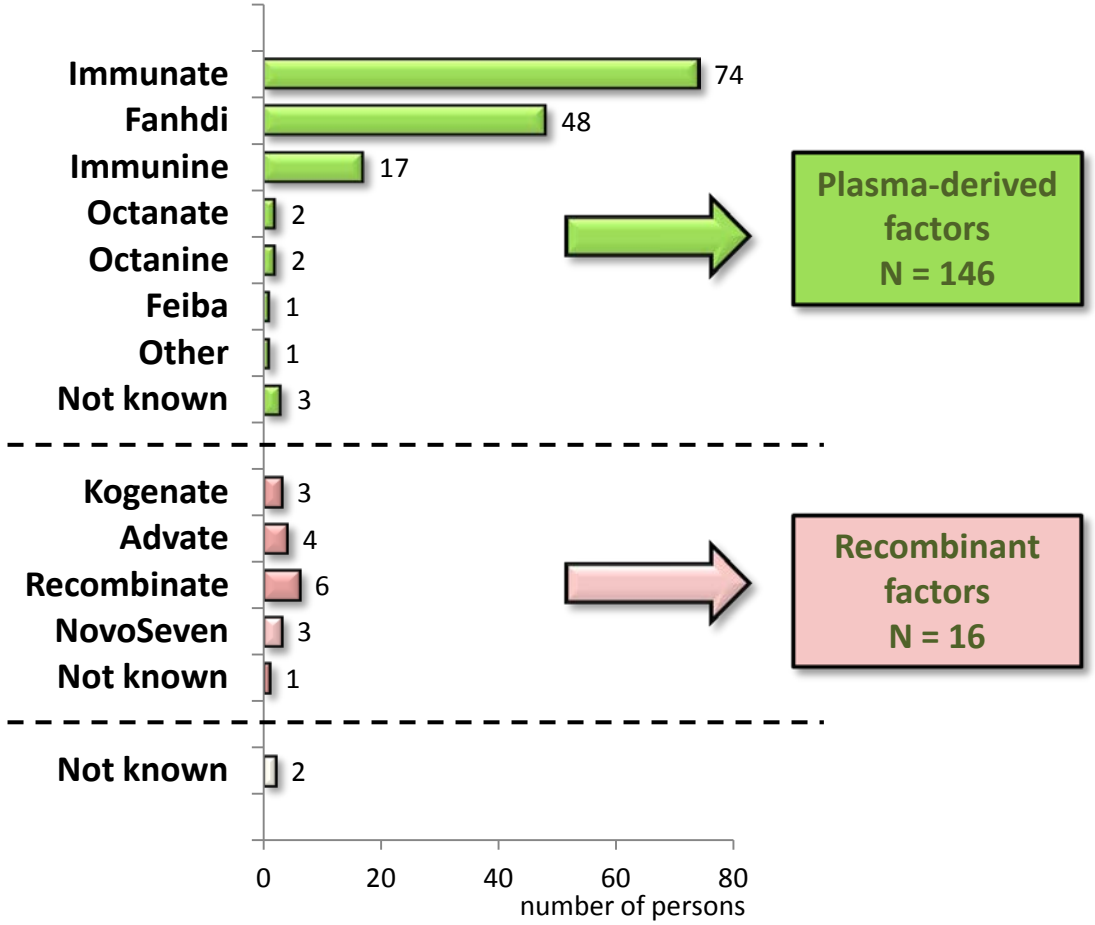


109 (64.1 %) children were treated in 2011 (13 of them with more different factor concentrates).
Plasma-derived factors were administered more frequently – in 81 children, recombinant factors in 37 children and not specified drug in 2 children.
10 children were treated with both plasma-derived and recombinant factor.

61 (35.9 %) children had no treatment in 2011.

Adults
N=211

Treatment

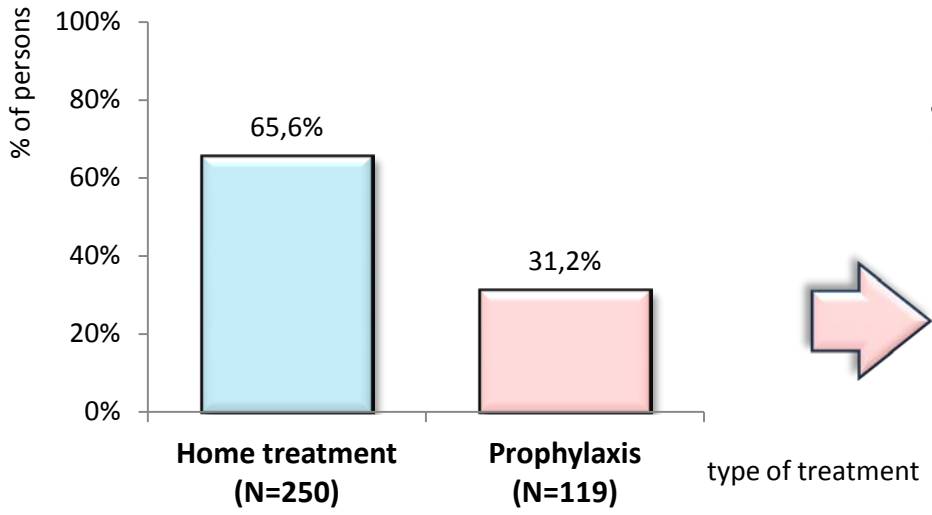


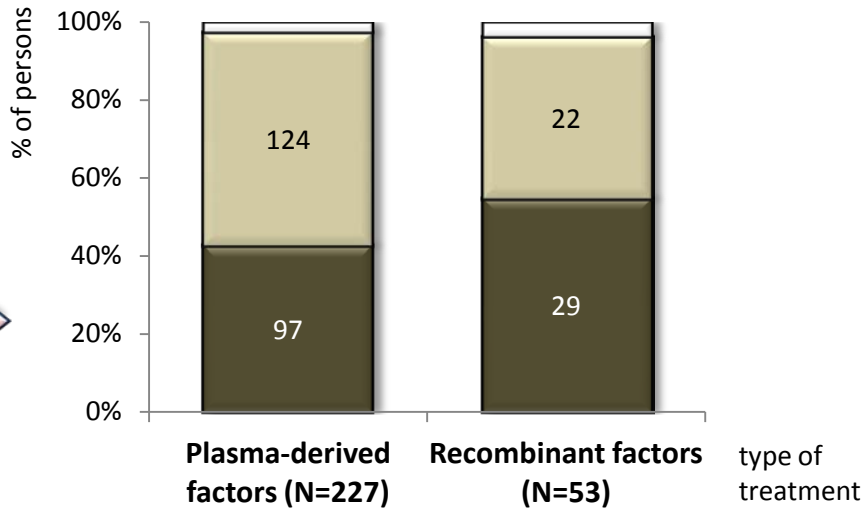
155 (73.5 %) adults were treated in 2011 (20 of them with more different factor concentrates).
Plasma-derived factors were administered more frequently – in 146 adults, recombinant factors in 16 adults and not specified drug in 2 adults.
9 adults were treated with both plasma-derived and recombinant factor.

56 (26.5 %) adults had no treatment in 2011.

All
N=381

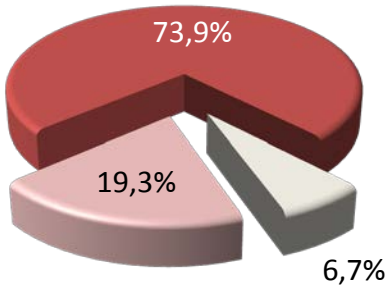
Type of treatment





Type of prophylaxis (N=119)

- Temporary (N=23)
- Permanent (N=88)
- Not available (N=8)



Prophylaxis

- Not available
- No
- Yes

Consumption of factors

All
N=381

	Factor concentrate	Total annual consumption	Number of treated persons	Average annual consumption per treated person	Number of examined persons	Average annual consumption per examined person
FVIII	Immunate	8 248 760 IU	115	71 728.3 IU		21 650.3 IU
	Fanhdi	5 840 700 IU	61	95 749.2 IU		15 329.9 IU
	Octanate	1 280 500 IU	12	106 708.3 IU		3 360.9 IU
	Other plasma-derived	291 500 IU	5	58 300.0 IU		765.1 IU
	Kogenate	1 408 500 IU	25	56 340.0 IU		3 696.9 IU
	Advate	1 050 250 IU	14	75 017.9 IU		2 756.6 IU
	Recombinate	763 500 IU	9	84 833.3 IU		2 003.9 IU
	Other recombinant	8 500 IU	2	4 250.0 IU		22.3 IU
	Other	48 800 IU	3	16 266.7 IU		128.1 IU
	FVIII celkem	18 941 010 IU	246	76 996.0 IU	381	49 713.9 IU
FIX	Immunine	1 928 800 IU	26	74 184.6 IU		5 062.5 IU
	Octanine	377 000 IU	6	62 833.3 IU		989.5 IU
	Other plasma-derived	49 200 IU	2	24 600.0 IU		129.1 IU
	FIX celkem	2 355 000 IU	34	69 264.7 IU		6 181.1 IU
aPCC	Feiba	2 000 IU	1	2 000.0 IU		5.2 mg
rFVIIa	NovoSeven	55.2 mg	3	18.4 mg		0.1 IU
Plasma-derived factors - TOTAL*		18 018 460 IU	227	79 376.5 IU		47 292.5 IU
Recombinant factors - TOTAL*		3 230 760 IU	48	67 307.3 IU		8 479.7 IU
TOTAL CONSUMTION *		21 298 010 IU	281	75 793.6 IU		55 900.3 IU

- plasma-derived factors = Immunate, Fanhdi, Octanate, Immunine, Octanine, Feiba, Other plasma-derived
- recombinant factors = Kogenate, Advate, Recombinate, Other recombinant

*TOTAL CONSUMPTION = all mentioned drugs excluding NovoSeven

Consumption of factors

Children
N=170

	Factor concentrate	Total annual consumption	Number of treated persons	Average annual consumption per treated person	Number of examined persons	Average annual consumption per examined person
FVIII	Immunate	2 545 750 IU	42	60 613.1 IU		14 975.0 IU
	Fanhdi	1 408 200 IU	14	100 585.7 IU		8 283.5 IU
	Octanate	1 190 500 IU	10	119 050.0 IU		7 002.9 IU
	Other plasma-derived	189 000 IU	3	63 000.0 IU		1 111.8 IU
	Kogenate	1 040 500 IU	22	47 295.5 IU		6 120.6 IU
	Advate	802 250 IU	11	72 931.8 IU		4 719.1 IU
	Recombinate	405 000 IU	4	101 250.0 IU		2 382.4 IU
	Other recombinant	2 500 IU	1	2 500.0 IU		14.7 IU
	Other	43 800 IU	2	21 900.0 IU		257.6 IU
	FVIII celkem	7 627 500 IU	109	69 977.1 IU	170	44 867.6 IU
FIX	Immunine	747 000 IU	10	74 700.0 IU		4 394.1 IU
	Octanine	116 000 IU	4	29 000.0 IU		682.4 IU
	Other plasma-derived	-	-	-		-
	FIX celkem	863 000 IU	14	61 642.9 IU		5 076.5 IU
aPCC	Feiba	-	-	-		-
rFVIIa	NovoSeven	-	-	-		-
Plasma-derived factors - TOTAL*		6 196 450 IU	81	76 499.4 IU		36 449.7 IU
Recombinant factors - TOTAL*		2 250 250 IU	37	60 817.6 IU		13 236.8 IU
TOTAL CONSUMTION *		8 490 500 IU	123	69 028.5 IU		49 944.1 IU

- plasma-derived factors = Immunate, Fanhdi, Octanate, Immunine, Octanine, Feiba, Other plasma-derived
- recombinant factors = Kogenate, Advate, Recombinate, Other recombinant

*TOTAL CONSUMPTION = all mentioned drugs excluding NovoSeven

Consumption of factors

Adults
N=211

	Factor concentrate	Total annual consumption	Number of treated persons	Average annual consumption per treated person	Number of examined persons	Average annual consumption per examined person
FVIII	Immunate	5 703 010 IU	73	78 123.4 IU	211	27 028.5 IU
	Fanhdi	4 432 500 IU	47	94 308.5 IU		21 007.1 IU
	Octanate	90 000 IU	2	45 000.0 IU		426.5 IU
	Other plasma-derived	102 500 IU	2	51 250.0 IU		485.8 IU
	Kogenate	368 000 IU	3	122 666.7 IU		1 744.1 IU
	Advate	248 000 IU	3	82 666.7 IU		1 175.4 IU
	Recombinate	358 500 IU	5	71 700.0 IU		1 699.1 IU
	Other recombinant	6 000 IU	1	6 000.0 IU		28.4 IU
	Other	5 000 IU	1	5 000.0 IU		23.7 IU
	FVIII celkem	11 313 510 IU	137	82 580.4 IU		53 618.5 IU
FIX	Immunine	1 181 800 IU	16	73 862.5 IU		5 600.9 IU
	Octanine	261 000 IU	2	130 500.0 IU		1 237.0 IU
	Other plasma-derived	49 200 IU	2	24 600.0 IU		233.2 IU
	FIX celkem	1 492 000 IU	20	74 600.0 IU		7 071.1 IU
aPCC	Feiba	2 000 IU	1	2 000.0 IU		9.5 IU
rFVIIa	NovoSeven	55.2 mg	3	18.4 mg		0.3 mg
Plasma-derived factors - TOTAL*		11 822 010 IU	146	80 972.7 IU		56 028.5 IU
Recombinant factors - TOTAL*		980 500 IU	11	89 136.4 IU		4 646.9 IU
TOTAL CONSUMTION *		12 807 510 IU	158	81 060.2 IU		60 699.1 IU

- plasma-derived factors = Immunate, Fanhdi, Octanate, Immunine, Octanine, Feiba, Other plasma-derived
- recombinant factors = Kogenate, Advate, Recombinate, Other recombinant

*TOTAL CONSUMPTION = all mentioned drugs excluding NovoSeven