Systém centrové péče o hemofiliky

(a příbuzné diagnózy)

v ČR

Demografie hemofilie v ČR (2012)

- Asi 850 HA, 130 HB v ČR
  - 4/5 dospělých
- Medián věku dospělých hemofiliků 40 let, děti 10
  - 5% nad 65 let
    - Nutnost lékařské péče jako o jiné stárnoucí pacienty
- Hemofilici narození po roce 1997 jsou “zdraví”
  - Profylaxe, prevence krvácení
- Starší hemofilici potřebují péči, vč. **CHIRURGICKÉ**
  - Ortopedie, obecná chirurgie, traumatologie, fyzio

Spotřeba FVIII v ČR

- V roce 2012 nad 4 IU/capita (parametr WFH)
  - Slovensko 6,5 IU/capita
  - Maďarsko 7 IU/capita
  - Švédsko >8 IU/capita
- Na co se faktor spotřebuje?
  - Profylaxe u dětí a mladých dospělých
  - Léčba krvácení + operativa u dospělých
  - Imunotolerance (inhibitor)
- Median spotřeby na léčeného dospělého hemofilika
  - 90 000 IU FVIII (cca 900 000 Kč vykázaných ZP)
    - Rozptyl od desítek tisíc po několik (desítek) milionů (inhibitor) Kč

Systém péče v ČR (2012)

- Český národní hemofilický program (ČNHP)
  - www.cnhp.cz
- Ročně se jedná o cca ¾ mld Kč pro cca 1000 lidí
- Systém center (2 úrovně)
  - 3 Komplexní (CCC): ÚHKT, FN Motol a FN Brno
  - 7 Léčebných (HTC): Ostrava, Hradec Králové, Olomouc, Plzeň, Ústí n L., Č. Budějovice, Liberec
- Zásadní odlišnost center:
  - Ortopedie/chirurgie (vč. elektivní), inhibitory – CCC
  - Bez chir oborů to “nejde” , zejména u dospělých

Úhrada péče o hemofilii v ČR (2013)

- V rámci DRG ALFA (většina chir diagnóz/výkonů)
  - Hrazeno plně bez omezení
  - Chir dg HLAVNÍ, Hemofilie VEDLEJŠÍ !
  - Nebyl problém s úhradou za hospitalisace
  - Jistá omezení v ambulanci (?)
- V rámci DRG BETA/GAMA
  - Hrazeno plně do výše stropu (výjimečně na chir prac)
- MIMO ÚV (tzv. “balíčky”, např TEP)
  - Individuální jednání ZZ a ZP
  - Snaha od r 2014 eliminovat tento způsob úhrady

Úhrada péče o hemofilii v ČR (2014)

- Pouze jeden DRG systém úhrady – Zrušena ALFA/ BETA/GAMA (týká se i chir diagnóz/výkonů)
  - Hrazeno plně bez omezení, bez zastropování v ambulanci i na lůžku
  - Hemofilie již může být hlavní dg!
  - Není tedy v principu problém s úhradou pro žádné ZZ, nedohodne-li jeho vedení se ZP jinak
    - Potíže mohou být se zálohami
    - Režim odlišný od S léku (netřeba mít smlouvu jako na centové léky)
    - Vztahuje se i na TEP

Co musí zajišťovat (navíc) CCC?

- Operativu hemofiliků s běžnými chir. Potížemi (desítky případů ročně)
- Operativu traumat u hemofiliků (ojedinělé případy ročně)
- Péče o osoby s inhibitorem
  - Jak chirurgie vč TEP
  - Tak ITT (většinou ale nejen děti)
- TEP kolene a kyčle (**2**-5 případů ročně – všechny krvácivé stavy, nejen hemofilie)
- Jiné elektivní ortopedické a chirurgické výkony
- Prenatalní diagnostiku
- Péči sexuologa, psychologa
- RHB a konsultace dalších odborností….

Požadavky na CCC/HTC

- Budou upraveny věstníkem MZd v roce 2014!

Proč by mělo mít ZZ zájem být centrem?

- Vysoce odborně prestižní oblast
- Možnost získat/udržet erudici a praxi, kterou jinde nemají
- Nejedná se o "ztrátový" či "virtuální" podnik
  - ÚV umožnuje úhradu v dostatečné výši (antihemofilika činí odhadem 80-90% nákladů na veškerou péči o tyto pacienty)
- Jedná se o relativně malé počty pacientů!!!
  - Nutná dostatečná expertíza. Maximum 2-3 CCC v zemi!
- Možnost vědeckého a odborného růstu pro pracoviště i jednotlivce
  - PhD studia, habilitace a pod… i pro "nehematology"

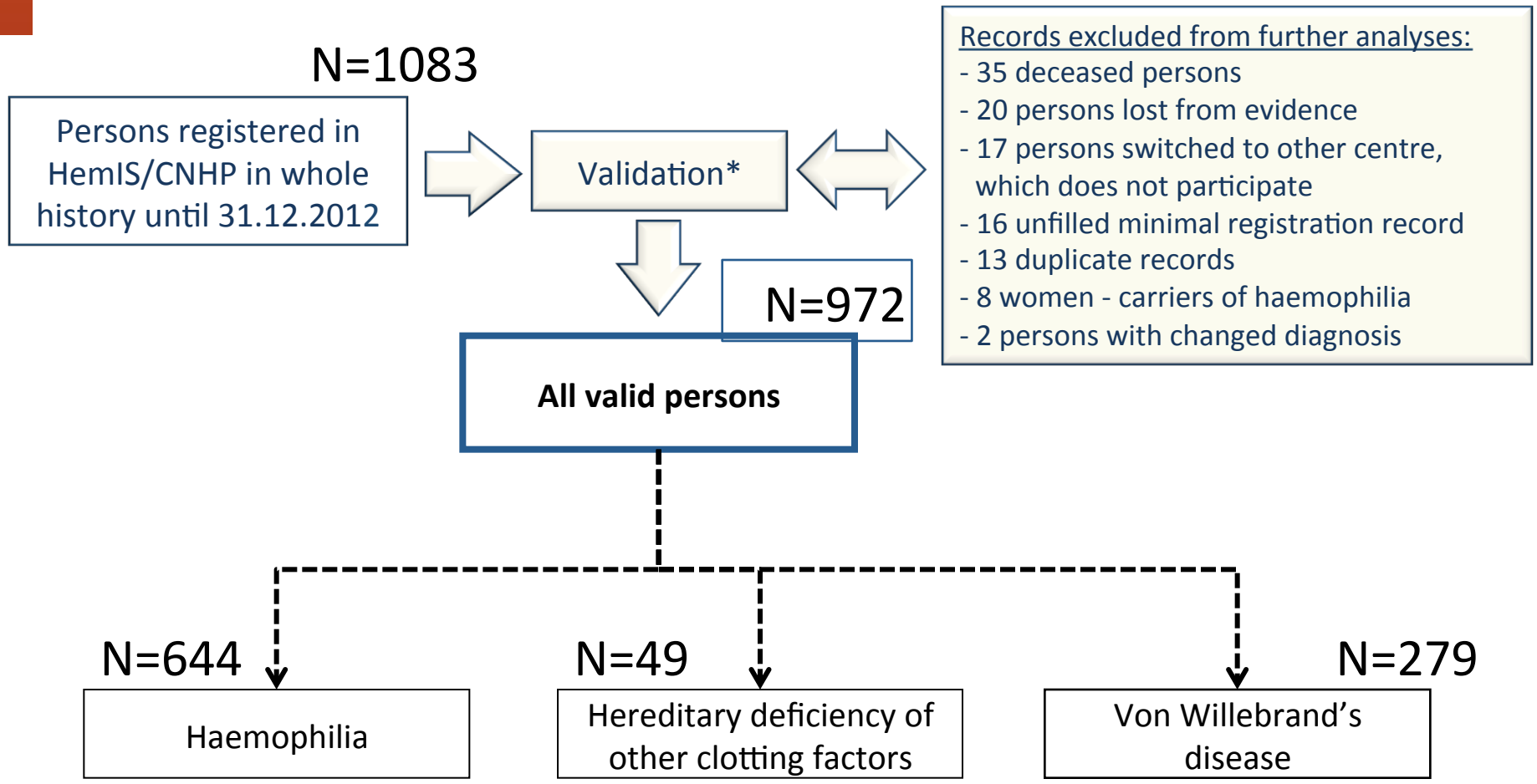
The current status of care for persons with haemophilia and von Willebrand's disease registered within CNHP registry

Jan Blatný, Petra Ovesná, Petr Brabec

on behalf of

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

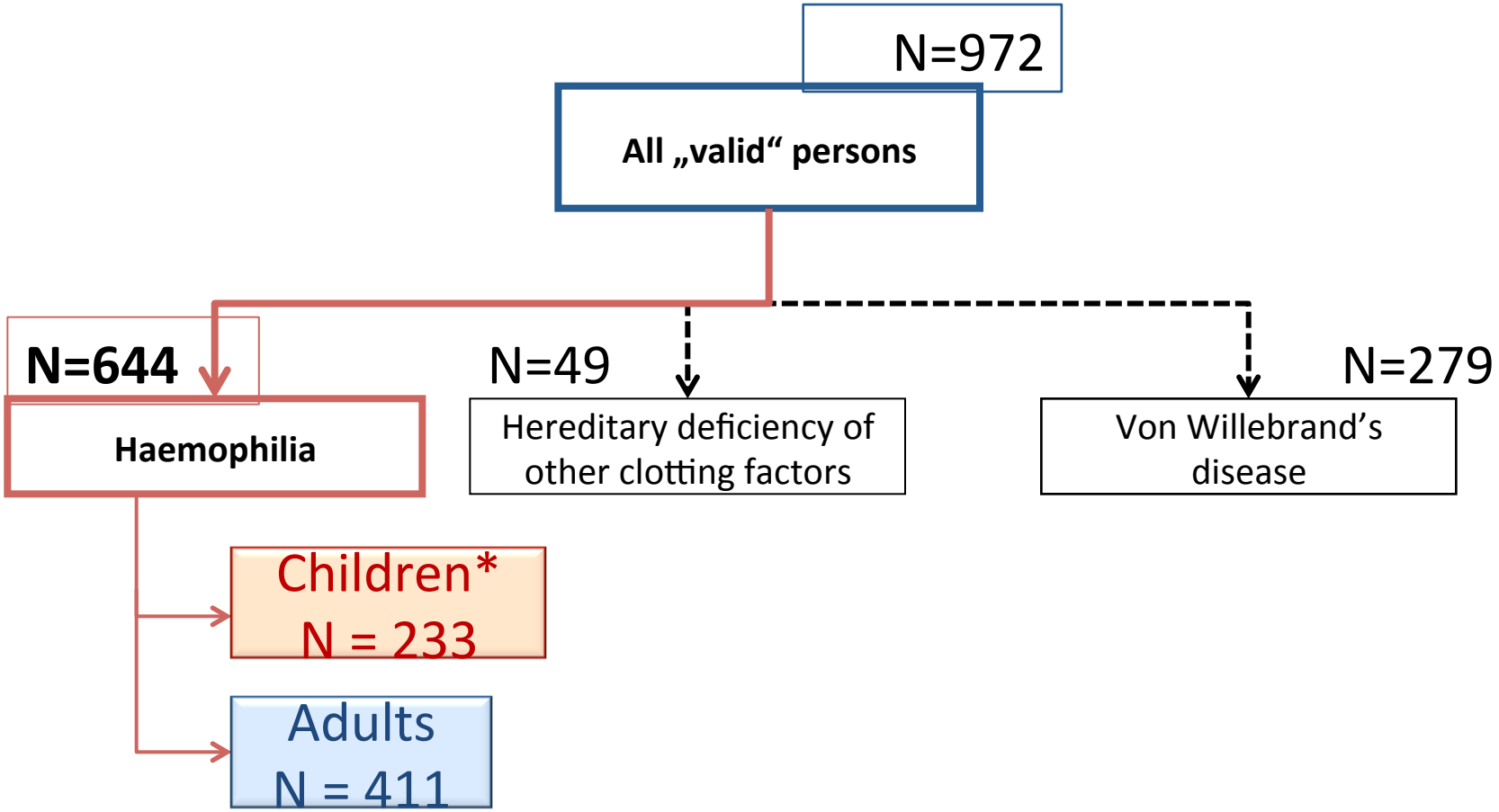
Sample size, valid records (2012)



Part A

Persons with haemophilia (PWH)

Sample size

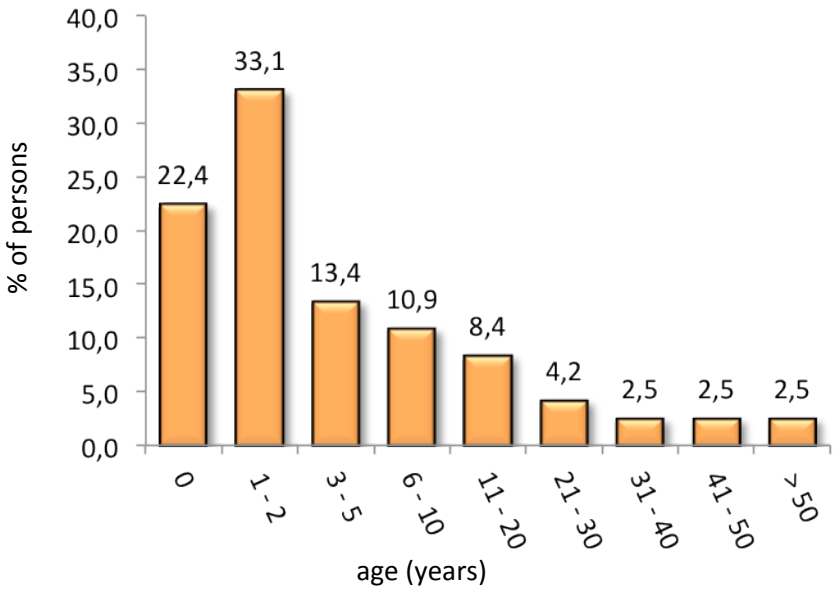


* *Persons under 19 years old in 2012*

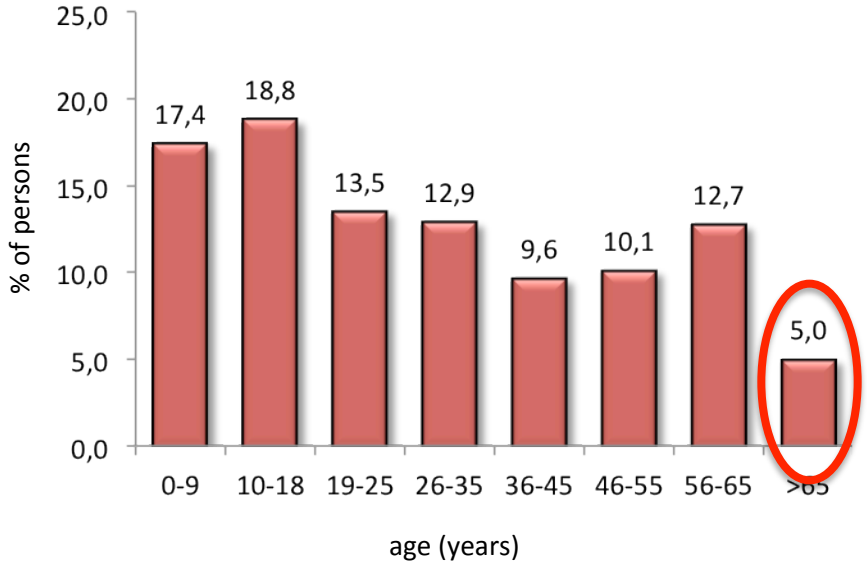
Age

All
N=644

	Age at diagnosis (years)
N	*477**
Mean	*7.6*
Median (min - max)	*2 (0 – 69)*



	Current age (years)
N	*644*
Mean	*30.5*
Median (min - max)	*26 (0 – 90)*

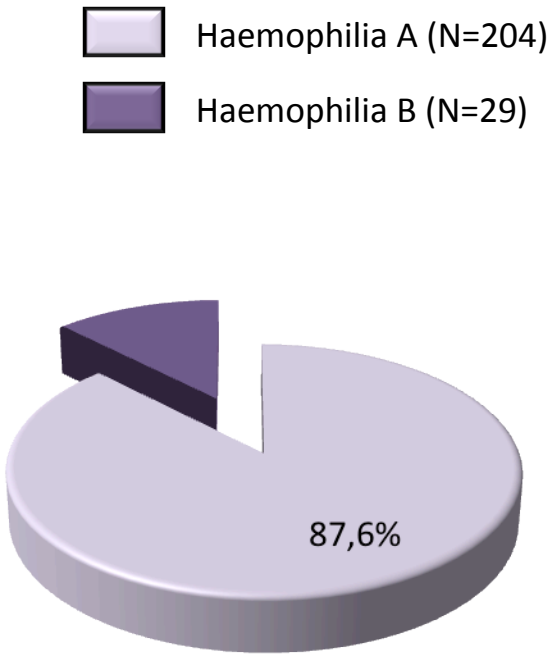


** Missing information on year of diagnosis in 167 persons.*

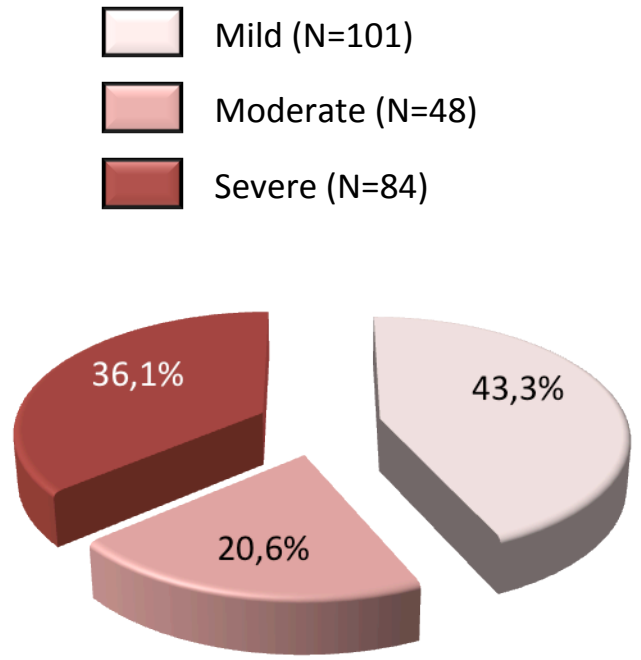
Type and severity of haemophilia I

Children
N=233

Type of haemophilia



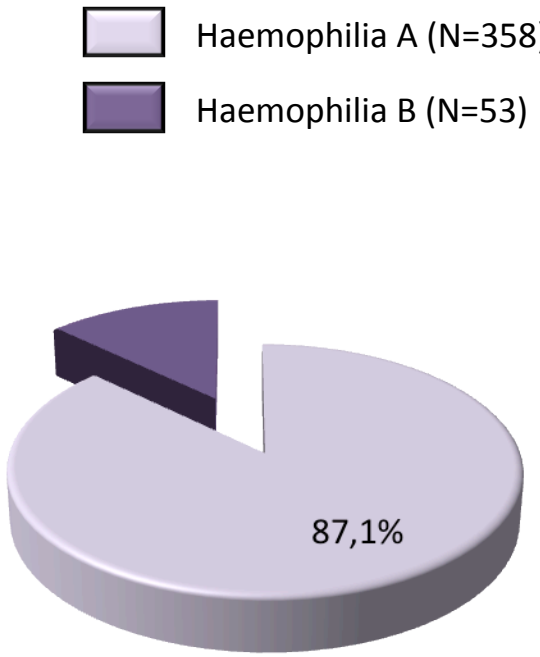
Severity of haemophilia (N=216)



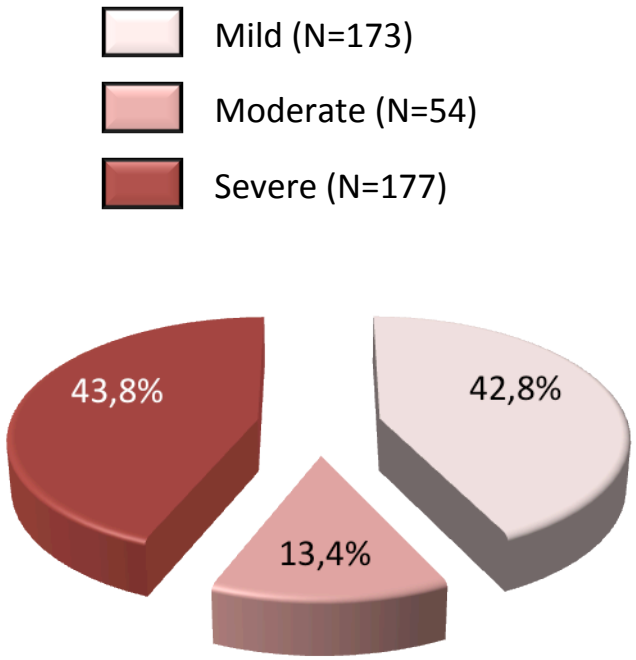
Type and severity of haemophilia I

Adults
N=411

Type of haemophilia



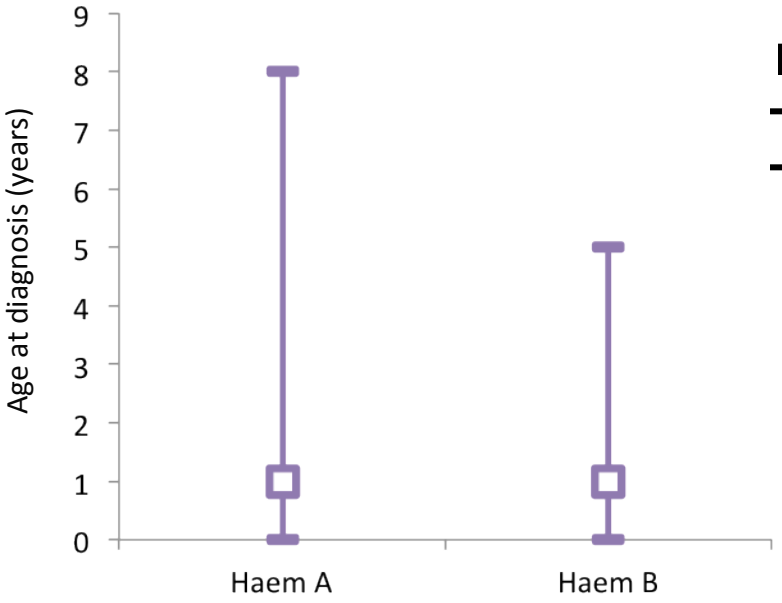
Severity of haemophilia (N=404*)

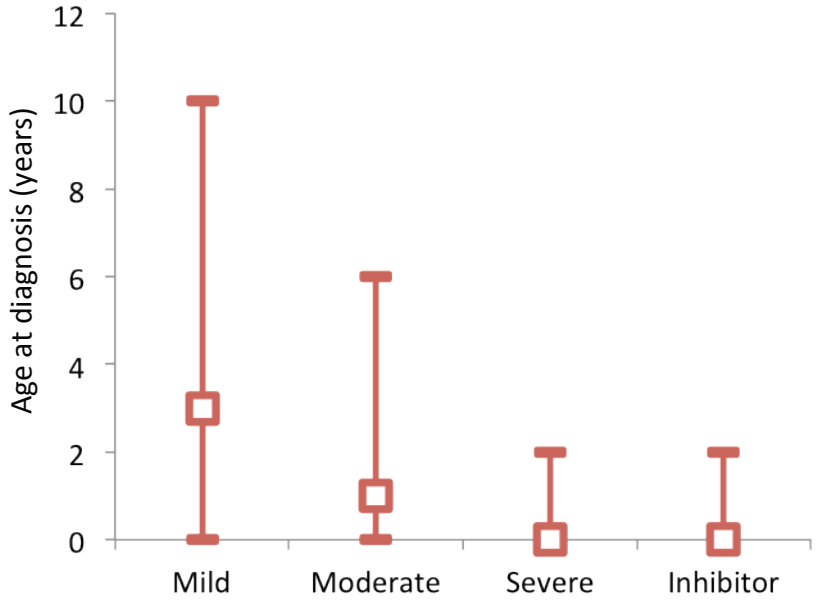


* *Severity of haemophilia not known in 7 adults.*

Age at diagnosis according to type and severity of haemophilia

Children
N=233



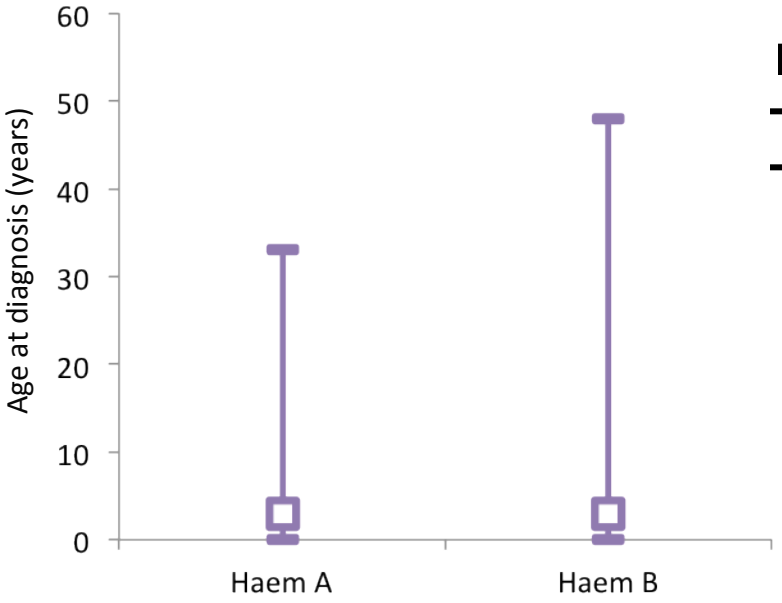


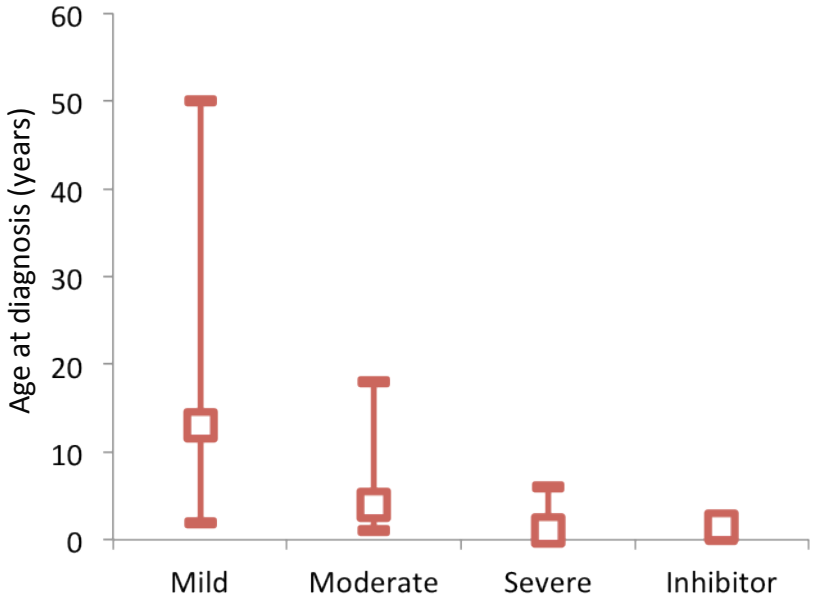
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
204	*29*	**N**	*101*	*48*	*84*	*5*
2.6	*2.3*	**Mean**	*4.0*	*2.6*	***0.9***	*0*
1 (0 – 17)	*1 (0 –13)*	**Median (min – max)**	*3 (0 – 13)*	*1 (0 – 17)*	***0*** *(0 – 11)*	*0 (0 – 2)*

** including persons with inhibitor*

Age at diagnosis according to type and severity of haemophilia

Adults
N=411



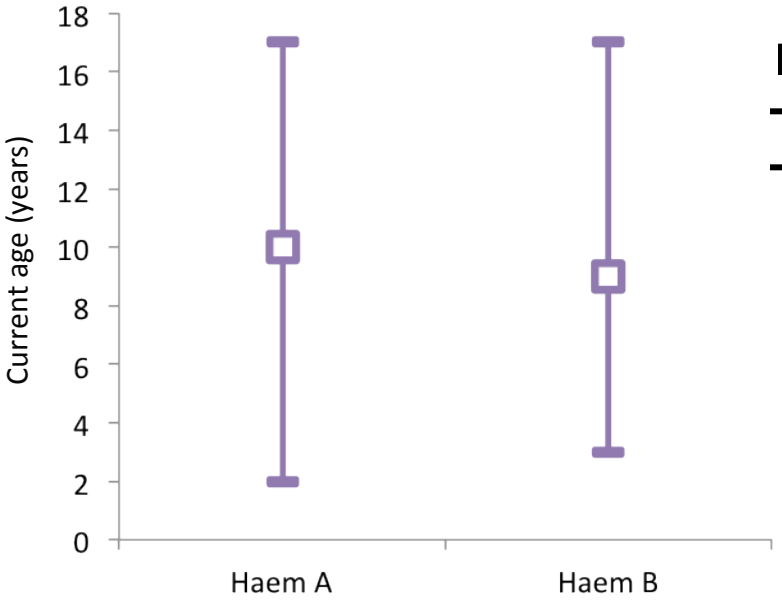


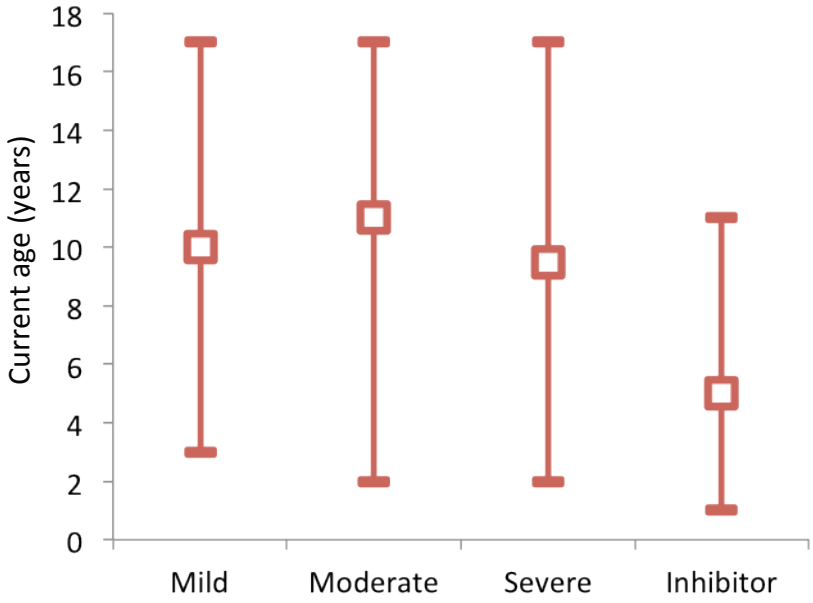
Haemophilia A	Haemophilia B	Age at diagnosis (years)	Mild*	Moderate*	Severe*	Inhibitor
358	*53*	**N**	*173*	*54*	*177*	*4*
10.9	*10.6*	**Mean**	*19.3*	*7.4*	*2.6*	*1.3*
3 (0 – 69)	*3 (0 – 63)*	**Median (min – max)**	*13 (1 – 69)*	*4 (0 – 61)*	*1 (0 – 48)*	*1.5 (0 – 2)*

** including persons with inhibitor*

Actual age according to type and severity of haemophilia

Children
N=233





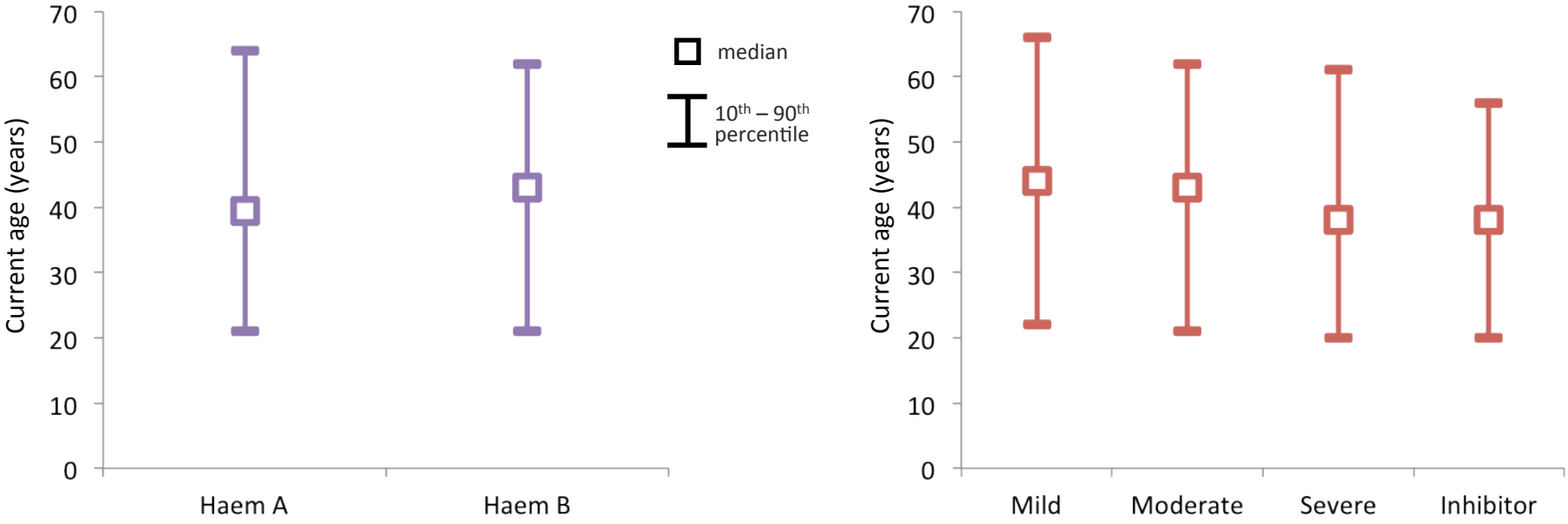
Haemophilia A	Haemophilia B	Current age* (years)	Mild*	Moderate*	Severe*	Inhibitor
204	*29*	**N**	*101*	*48*	*84*	*5*
9.8	*9.6*	**Mean**	***9.8***	***10.5***	***9.3***	***5.4***
10 (0 – 18)	*9 (2 – 18)*	**Median (min – max)**	*10 (0 – 18)*	*11 (0 – 18)*	*9.5 (0 – 18)*	*5 (1 – 11)*

**Current age = age reached in year 2012*

** including persons with inhibitor*

Actual age according to type and severity of haemophilia

Adults N=411



Haemophilia A	Haemophilia B	Current age* (years)	Mild*	Moderate*	Severe*	Inhibitor
358	*53*	**N**	*173*	*54*	*177*	*4*
42.3	*41.5*	**Mean**	***44.2***	***42.5***	***39.7***	***38.0***
39.5 (19 – 90)	*43 (20 – 67)*	**Median (min – max)**	*44 (19 – 90)*	*43 (19 – 71)*	*38 (19 – 73)*	*38 (20 – 56)*

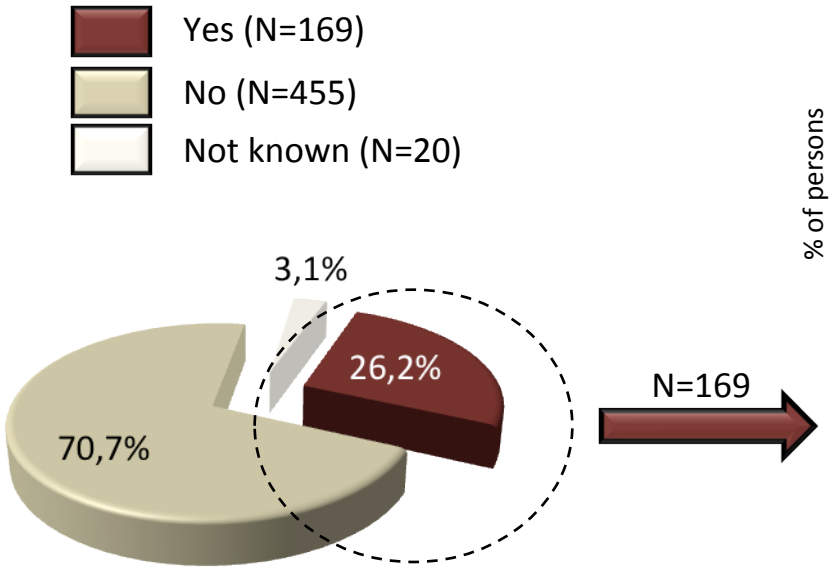
**Current age = age reached in year 2012*

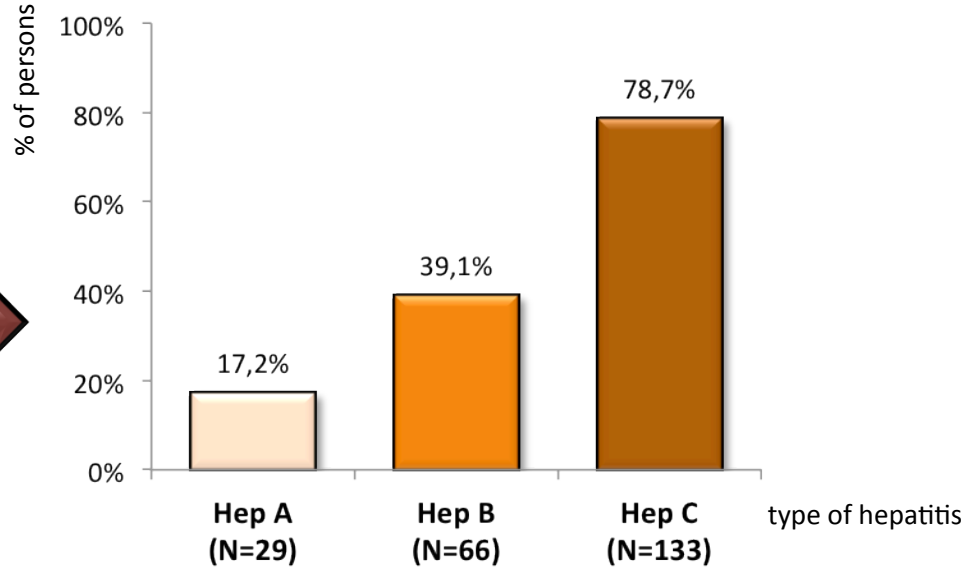
** including persons with inhibitor*

All
N=644

Hepatitis experienced

Experienced hepatitis





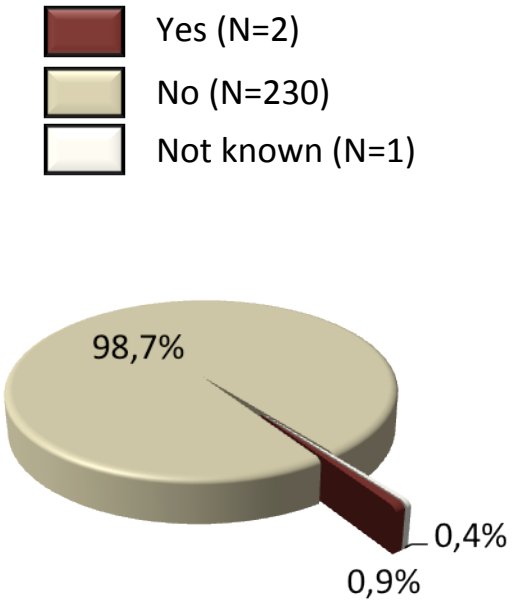
Data from last annual report of each person.

Type of hepatitis not specified in 5 persons. One person may have recorded more types of hepatitis.

Hepatitis experienced

Children
N=233

Experienced hepatitis



N=2

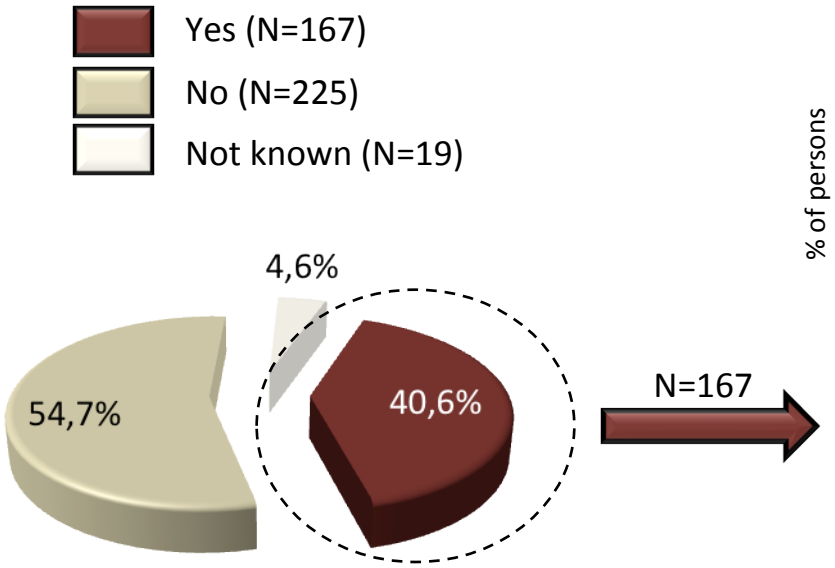
One child has hepatitis C, type of hepatitis not specified in one child.

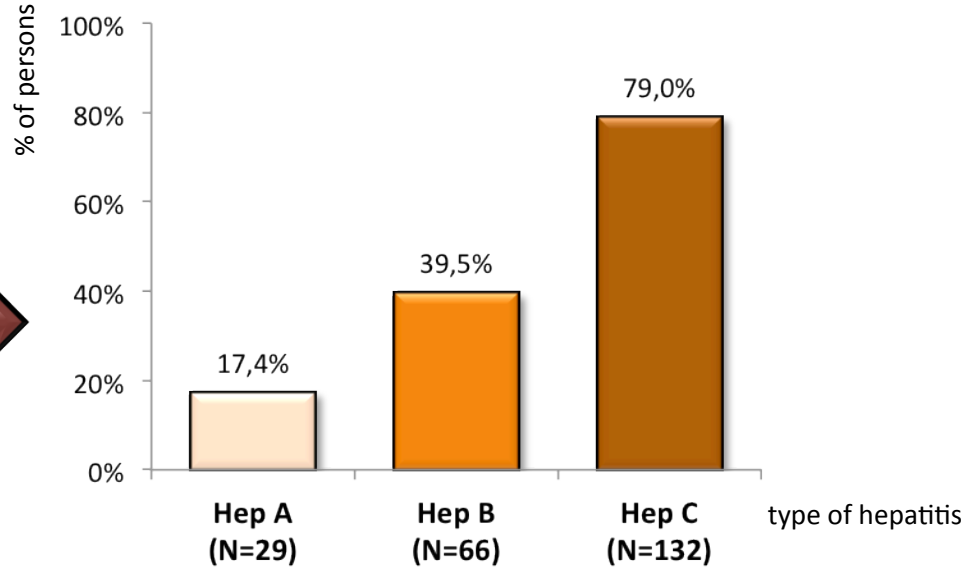
Data from last annual report of each person.

Adults
N=411

Hepatitis experienced

Experienced hepatitis





Data from last annual report of each person.

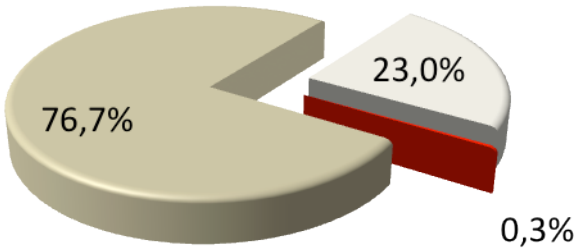
Type of hepatitis not specified in 3 adults. One person may have recorded more types of hepatitis.

All
N=644

HIV

HIV

Positive (N=2)
Negative (N=494)
Not known / not available (N=148)

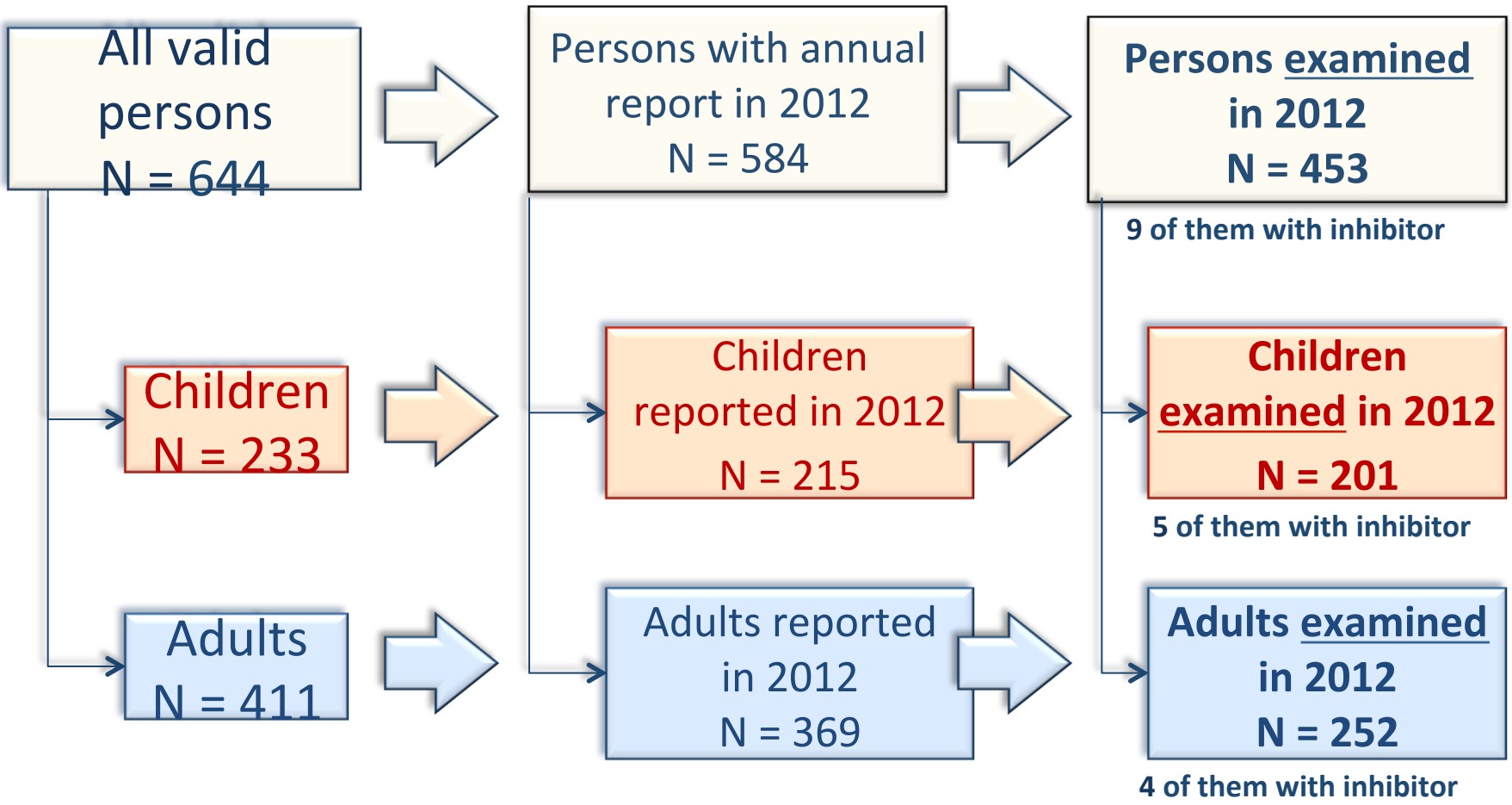


N=2

Both HIV-positive persons are adults.

Data from last annual report of each person.

Data from year 2012 – sample size



All
N=453

Persons with haemophilia with inhibitor

- inhibitor was recorded in 8 persons in year 2012
- other 1 person has recorded inhibitor in 2011 (data from 2012 are not available)

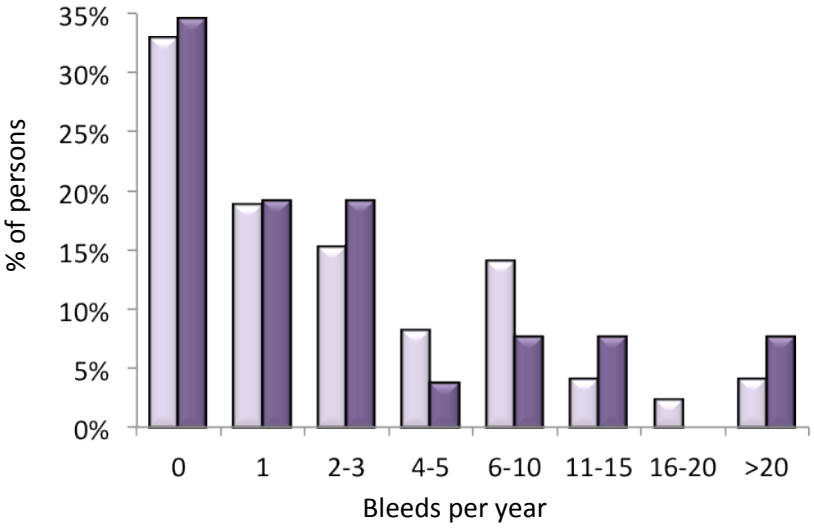
currently 9 persons with inhibitor

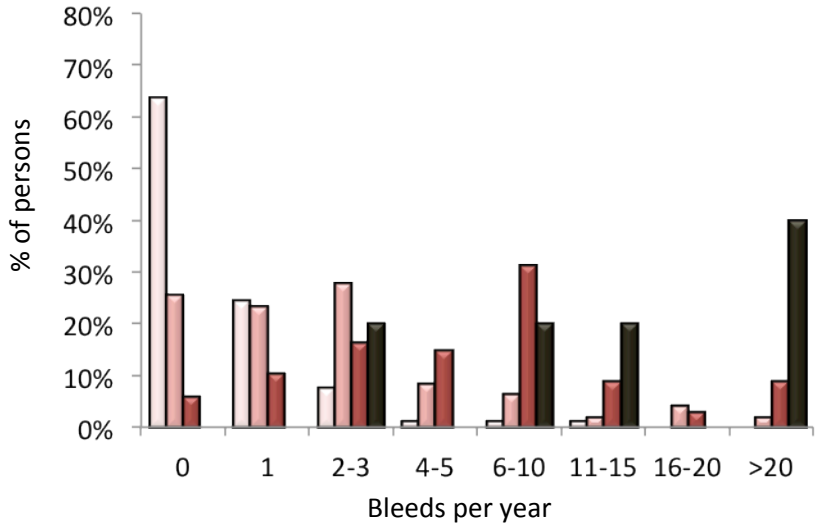
(5 children and 4 adults) + 6 in other centre

  - **NO increase in inhibitor incidence** rate in PUPs with Haemophilia A **since introduction of rFVIII** in 2003!
    - Annual incidence around 5%, absolute around 20% (submitted for publication 2014, EAHAD poster 090, 2014)

Frequency of bleeding requiring treatment in 2012

Children
N=201





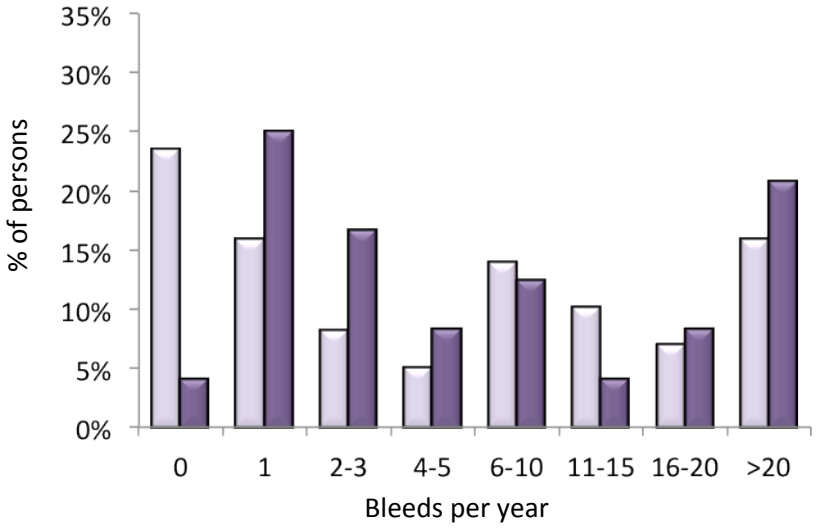
Haemophilia A	Haemophilia B	Frequency of bleeding	Mild*	Moderate*	Severe*	Inhibitor
175	*26*	**N total**	*79*	*47*	*70*	*5*
170	*26*	**N valid**	*77*	*47*	*67*	*5*
4.3	*4.4*	**Mean**	*0.8*	*3.6*	*8.0*	*15.2*
1 (0 – 49)	*1 (0 – 26)*	**Median (min – max)**	*0 (0 – 12)*	*2 (0 – 30)*	*6 (0 – 49)*	*11 (3 – 32)*

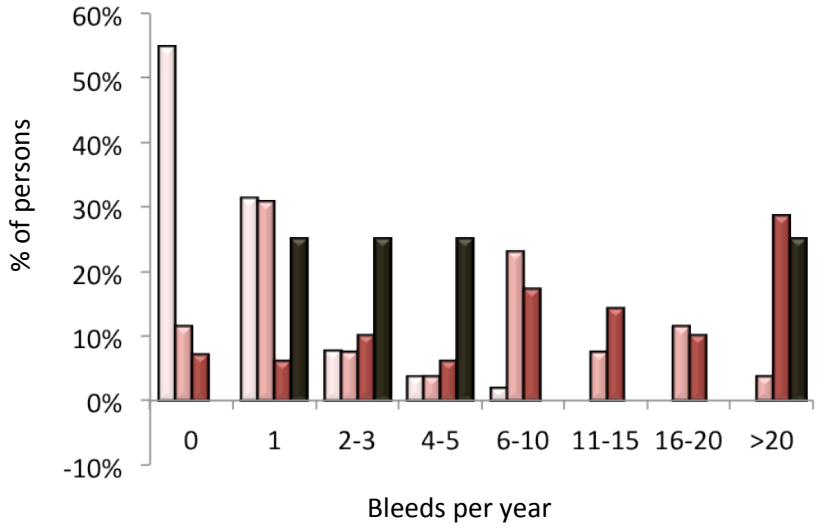
** without inhibitor*

Frequency of bleeding is missing in 19 children.

Frequency of bleeding requiring treatment in 2012

Adults N=252





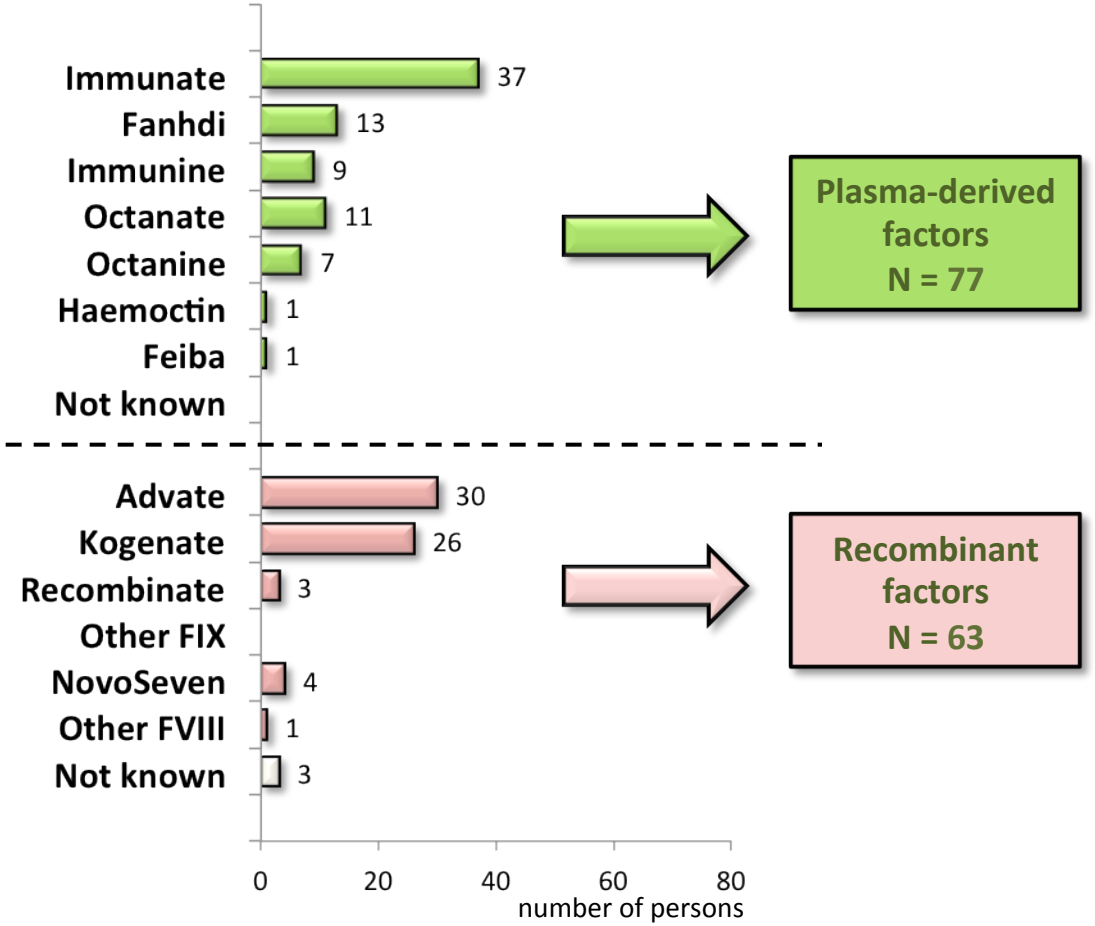
Haemophilia A	Haemophilia B	Frequency of bleeding	Mild*	Moderate*	Severe*	Inhibitor
211	*41*	**N total**	*64*	*36*	*144*	*4*
157	*24*	**N valid**	*51*	*26*	*98*	*4*
10.5	*15.3*	**Mean**	*0.8*	*6.5*	*17.7*	*8.5*
5 (0 – 144)	*5 (0 – 132)*	**Median (min – max)**	*0 (0 – 7)*	*3.5 (0 – 31)*	*12.5 (0 – 144)*	*3.5 (1 – 26)*

** without inhibitor*

Frequency of bleeding is missing in 188 adults.

Treatment

Children
N=201



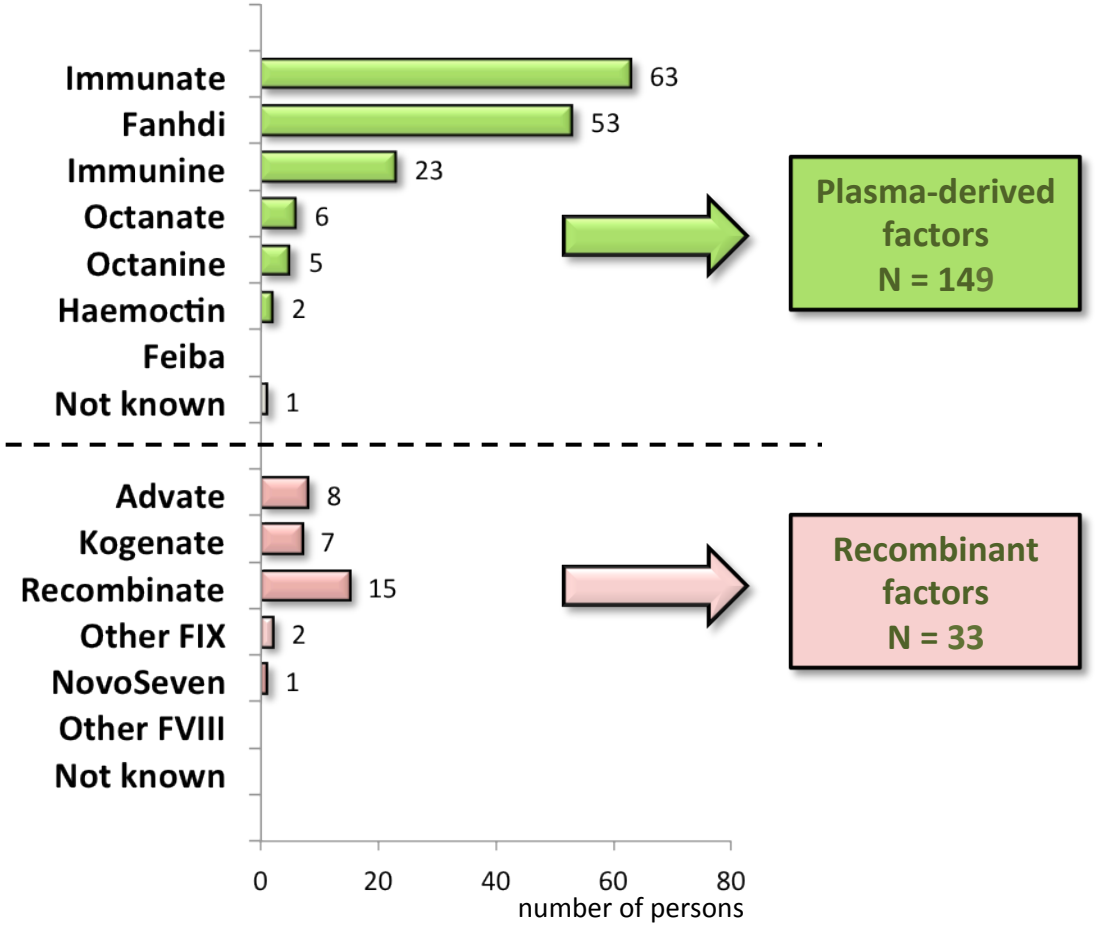
Plasma-derived factors
N = 77

Recombinant factors
N = 63

135 (67.2%) children received drugs in 2012 (11 of them more different drugs).
Plasma-derived factors were administered in 77 (38.3%) children, recombinant factors in 63 (31.3%).
Five children were treated with both plasma-derived and recombinant factor.

Treatment

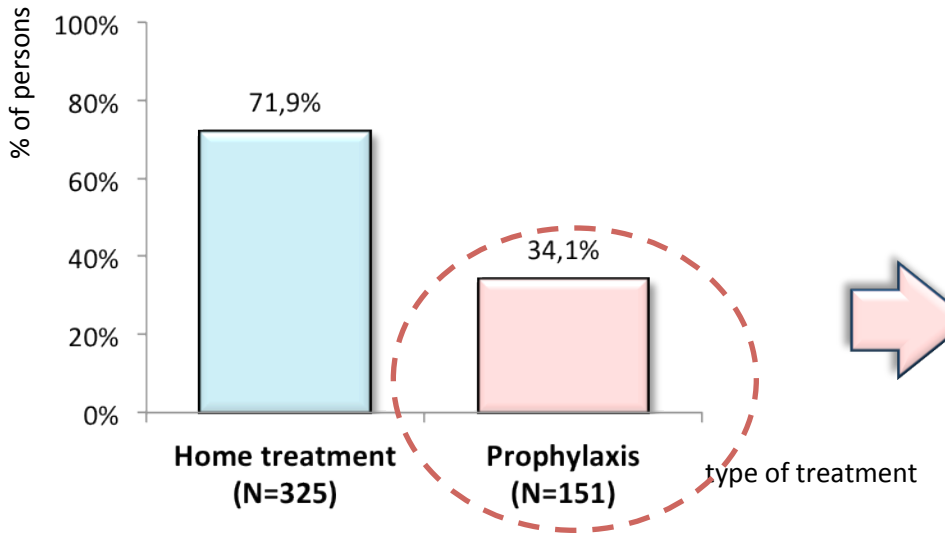
Adults
N=252

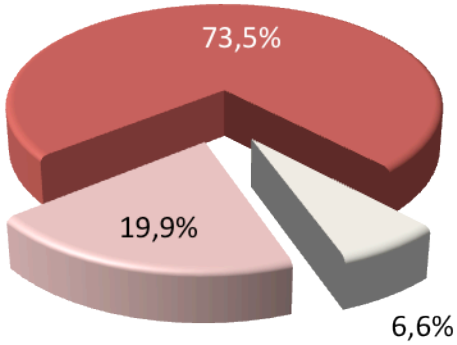


177 (70.2%) adults received drugs in 2012 (9 of them more different drugs).
Plasma-derived factors were administered more frequently – in 149 (59.1%) adults, whereas recombinant factors in 33 (13.1%) adults.
Five adults were treated with both plasma-derived and recombinant factor.

All
N=453

Type of treatment



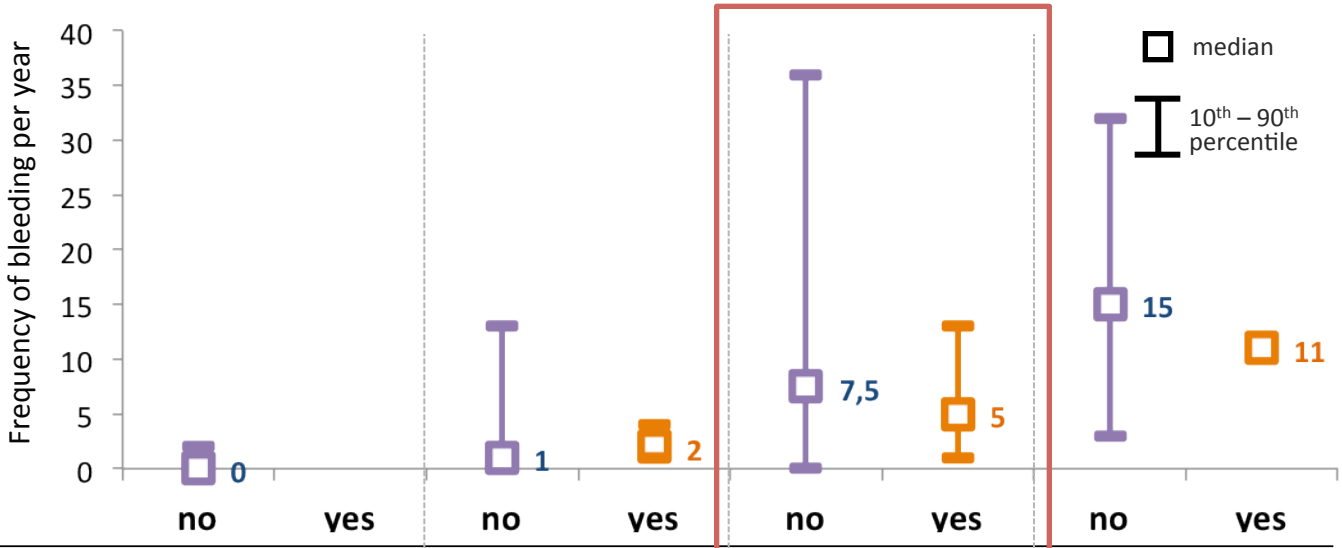


Type of prophylaxis (N=151)

- Temporary (N=30)
- Permanent (N=111)
- Not available (N=10)

Bleeding requiring treatment according to prophylaxis

Children
N=201



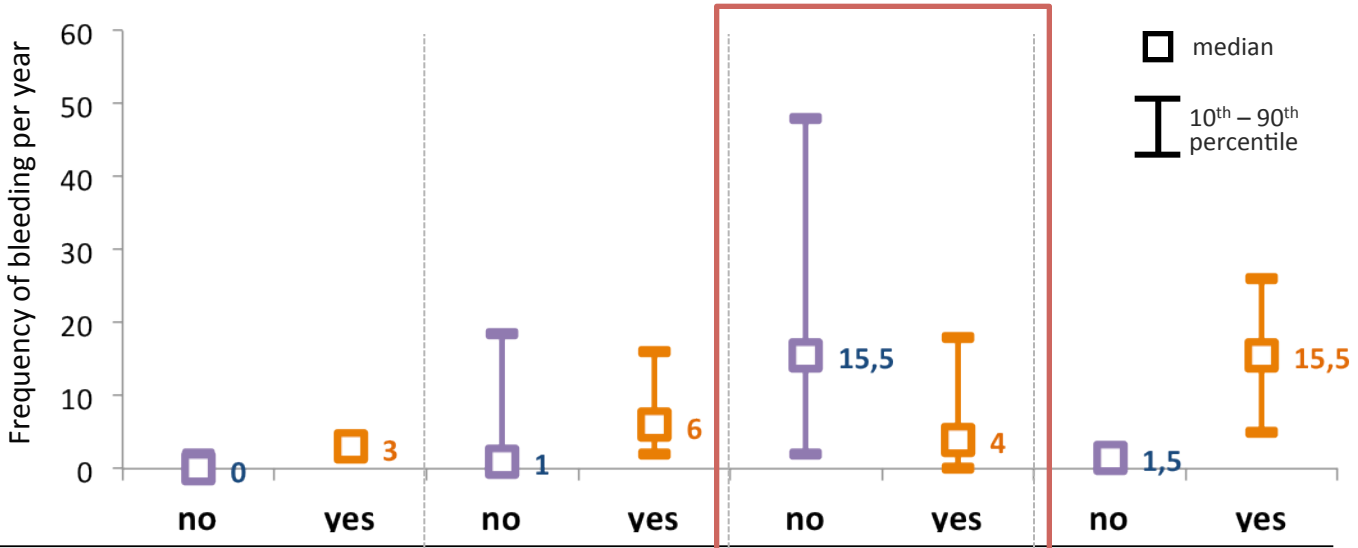
permanent prophylaxis

Frequency of bleeding	Mild*	Moderate*		Severe*		Inhibitor	
N	*77*	*38*	*9*	*18*	*49*	*4*	*1*
Mean	*0.8*	*3.9*	*2.2*	***13.2***	***6.1***	*16.3*	*11.0*
Median (min – max)	*0 (0 – 12)*	*1 (0 – 30)*	*2 (1 – 4)*	***7.5 (0 – 49)***	***5 (0 – 25)***	*15 (3 – 32)*	*11*
children on permanent prophylaxis	***0 (0%)***	***9 (19.1%)***		***51 (72.9%)***		***1 (20%)***	
% of drugs (FVIII and FIX) consumed by children on permanent prophylaxix	***0 %***	***9 %***		***70 %***		***4 %***	

** without inhibitor*

Bleeding requiring treatment according to prophylaxis

Adults
N=252



Frequency of bleeding	Mild*		Moderate*		Severe*		Inhibitor	
N	50	1	20	6	68	30	2	2
Mean	0.8	3.0	6.3	7.5	22.8	6.3	1.5	15.5
Median (min – max)	0 (0 – 7)	3	1 (0 – 31)	6 (2 – 16)	15.5 (0–144)	4 (0 – 31)	1.5 (1 – 2)	15.5 (5 – 26)
adults on permanent prophylaxis	1 (1.7%)		8 (22.9%)		38 (27%)		2 (50%)	
% of drugs (FVIII and FIX) consumed by adults on permanent prophylaxix	1.4 %		6.5 %		42.8 %		0.9 %	

* without inhibitor

Děkuji za pozornost!!