

Twenty-two years of modern haemophilia care in The Czech Republic

Jan Blatný, Petr Smejkal, Vladimír Komrska, Zuzana Čermáková, Hana Płoszková, Petr Dulíček, Jiří Hak, Daniela Procházková, Zdeňka Černá, Dagmar Pospíšilová, Antonín Hluší, Pavel Timr, Ivan Vonke, Zdeňka Hajšmanová,, Lenka Walterová, Petr Kessler, Miroslav Penka on behalf of Czech National Haemophilia Programme (CNHP).

Before 1989

Czech Republic with its 79 000 km² and over 10 million inhabitants is a home country for over 900 people with haemophilia. Before year 1989 the “iron curtain” was shut down and factor concentrates were in fact not available in the former Czechoslovakia. Haemophilia care was almost solely based on the treatment with FFP and cryoprecipitate. We could only dream about home treatment and prophylaxis. The only “benefit” of the country isolation at that time was the low incidence of HIV infected people, including haemophiliacs.



After “Velvet revolution”

Since 1990 the concentrates become available also for Czech persons with haemophilia (PWH). In accord with WFH recommendations and later on with the European principles of haemophilia care ⁽¹⁾ the network of haemophilia centres (CCC, HTC) was built up and anchored firmly within The Czech National Haemophilia Programme (CNHP).



The “Run for haemophilia” could finally go on...

We started with proper and efficient “on-demand” treatment and offered the advantage of “home treatment” to all people with haemophilia, who needed it. Further step was the introduction of short-term prophylaxis after significant bleeds, during physiotherapy and during/after surgical intervention. At that time we also started ambitious surgical intervention programme to get our patients “back from their wheelchairs”.

Late 90s

The availability and quality of factor concentrates further improved. Medium to high purity plasma derived concentrates became available and we were able to introduce the major change for the Czech haemophilia population – the long-term prophylaxis for all children, who needed it. We started with the secondary prophylaxis, which was, at that time, close to so called “Dutch regimen” using lower doses (in median 24 IU/kg 2–3 times per week) We tailored the treatment to the needs of particular child being led not solely by the genotype, but rather by the phenotype of the disease. Thus we were able to cover all children with proper treatment for affordable costs. Summer Haemophilia Camps, where children learn the self-administration of their concentrates were introduced and since that time they play important role in patients’ education. We continued in building up the sophisticated haemophilia centres network.

Conclusion

Authors believe, that during past 22 years the Czech haemophilia care has been built up to the level comparable with other European countries where this care has long tradition. We also want to emphasize that during relatively short time it is possible to significantly improve the care of PWH for reasonable and affordable costs. We understand, that our “run for haemophilia” is not over and that there are many “Czechllenges” we will have to face in the future.

Acknowledgements

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We also want to thank IBA (Institute of Biostatistics and Analyses) of Masaryk University in Brno for technical support within CNHP.

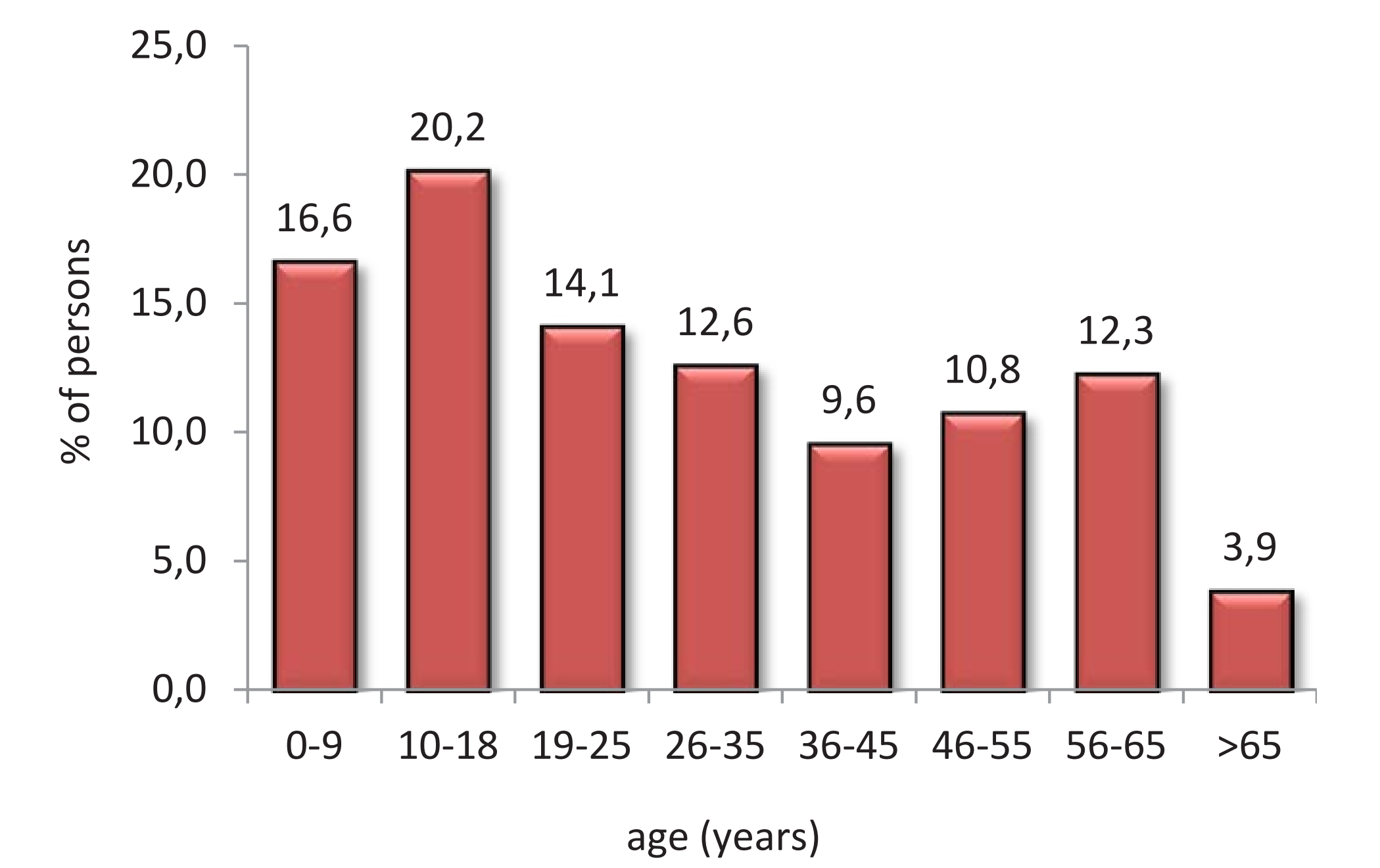
Data from 2011 Annual CNHP report have been used for this presentation.

Five pillars of the “Bridge over troubled water” our run for haemophilia went over

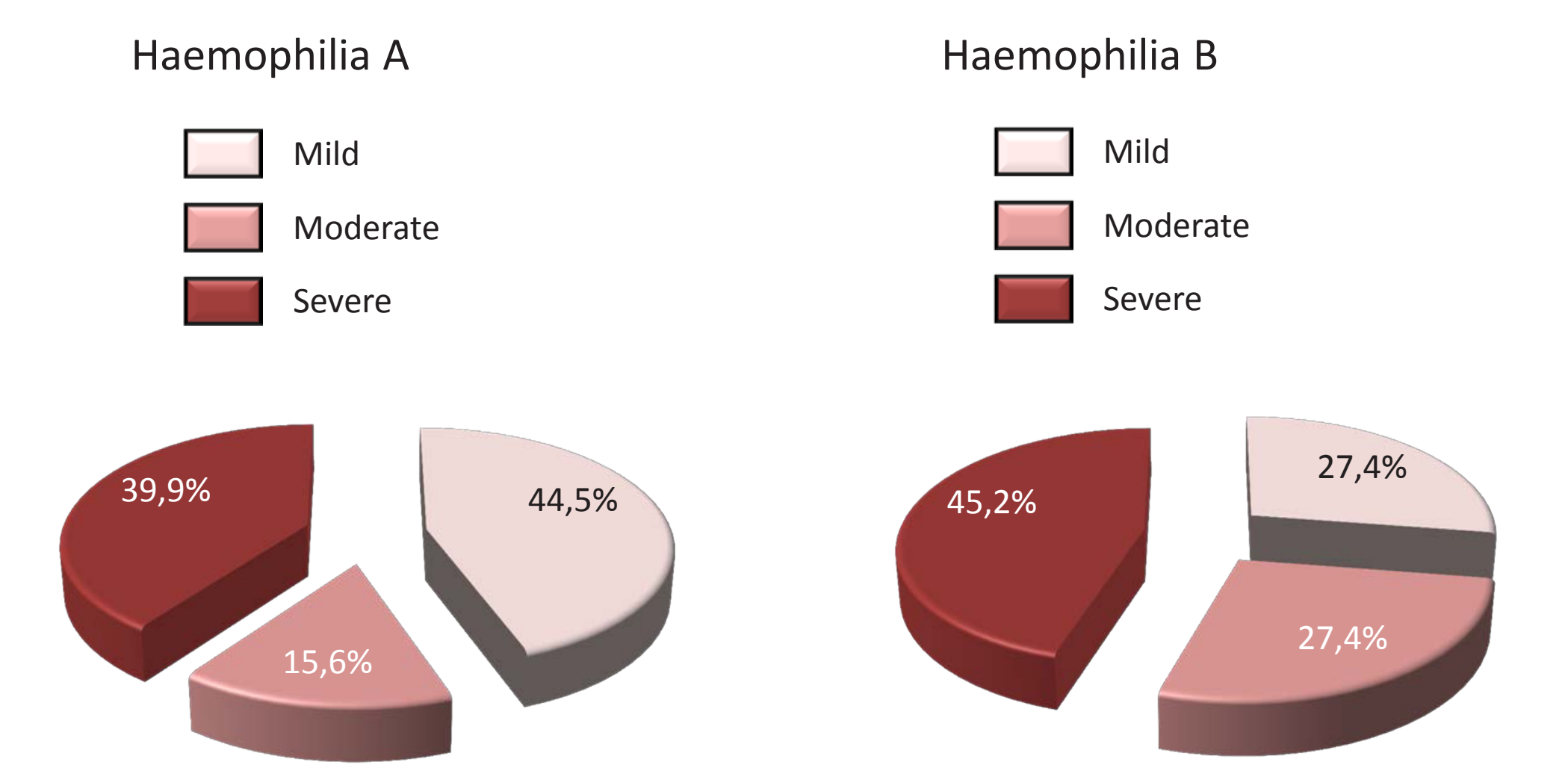
- Prophylaxis to ALL kids with severe phenotype
- Home treatment available for ALL
- Keep maximal safety of the treatment
 - “iron curtain” was uncrossable also for blood born infections including HIV!!!
 - Only around 30 PWH (including 7 children) infected with HIV
- Efficient and immediate treatment for patients with inhibitors
- Building up the Haemophilia Centres network

Half way behind us

The demography of Czech haemophilia population is shown on graphs 1 and 2.

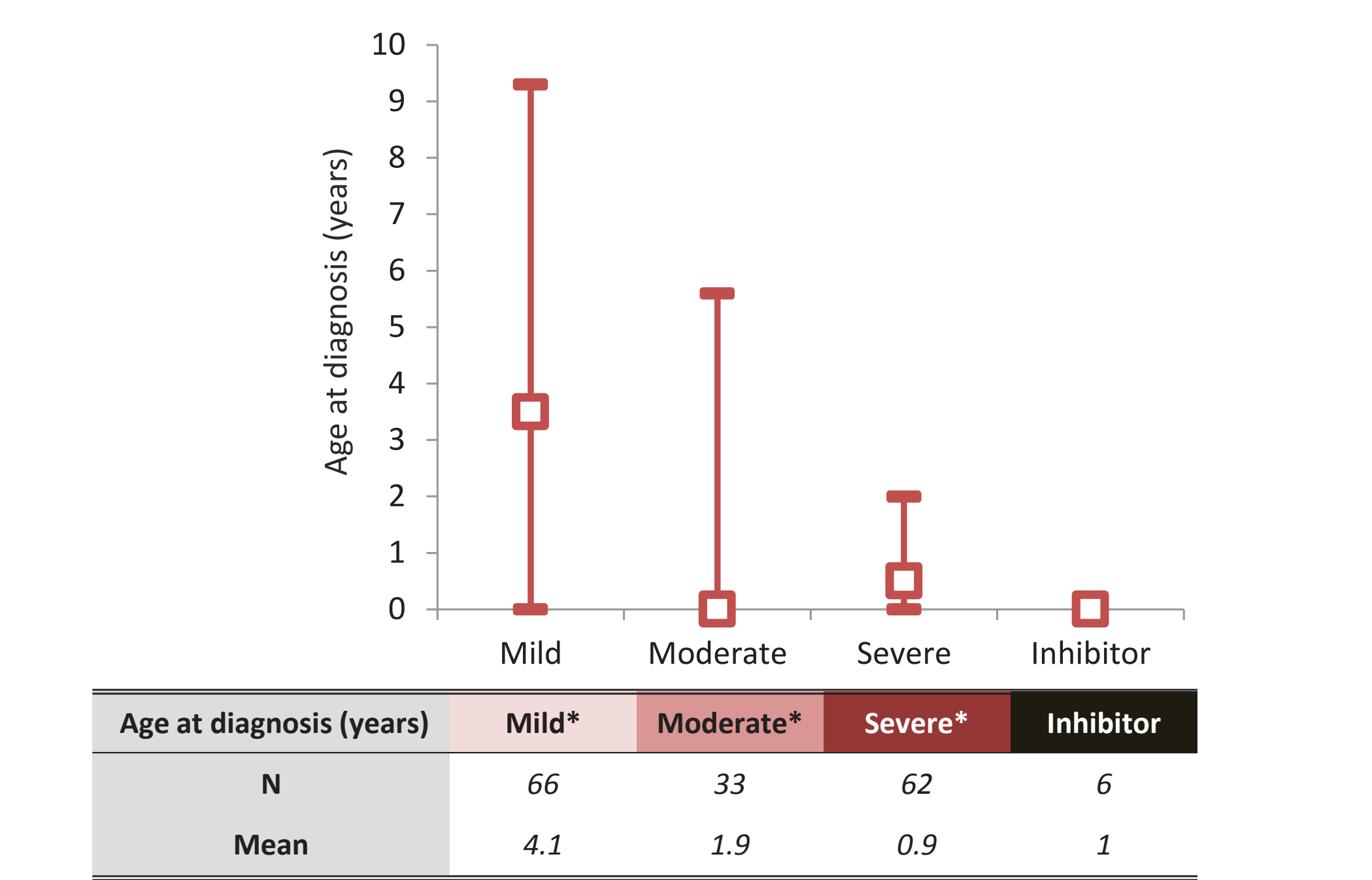


Graph 1. Age of Czech Persons with Haemophilia (PWH)



Graph 2. Type and severity of Haemophilia in Czech Republic

As shown on graph 3, we are currently able to confirm the diagnosis of severe and moderate haemophilia in our children before 2 years of age and thus we are able to offer them the primary prophylaxis regimens.



Graph 3. Age at diagnosis in Czech children with haemophilia

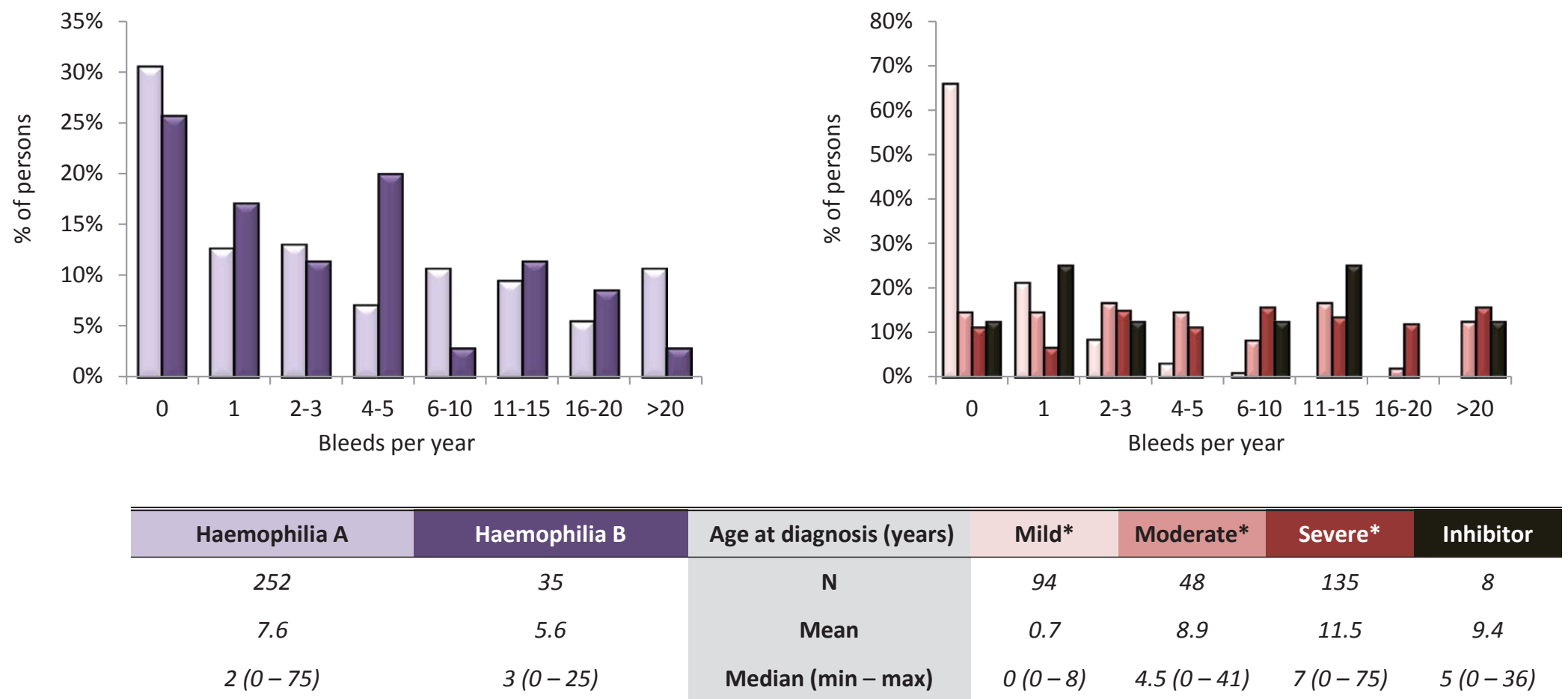
Since 2006 in accord with Czech national recommendations the treatment of choice for newly diagnosed severe haemophiliacs is primary prophylaxis. Currently we are using the regimen described by Auerswald, Kurnik et al. ⁽²⁾. Young adults are encouraged to continue on prophylaxis started during their childhood. For newly diagnosed haemophilia A children we recommend rFVIII as a first line treatment. Recombinant FIX is currently not available on Czech market. PWH, who are not treated with recombinants are treated with high purity plasma derived products. Consumption of FVIII in Czech Republic is currently around 3.6 IU/capita. In median we used about 78 000 IU of FVIII per treated patient per year in our haemophilia centres in 2011. About 10 % of that consumption is covered by recombinants. This number, however, represents about 20 % of children with haemophilia being currently treated with recombinant concentrates and it is continuously increasing since 2006.

Safety and inhibitors

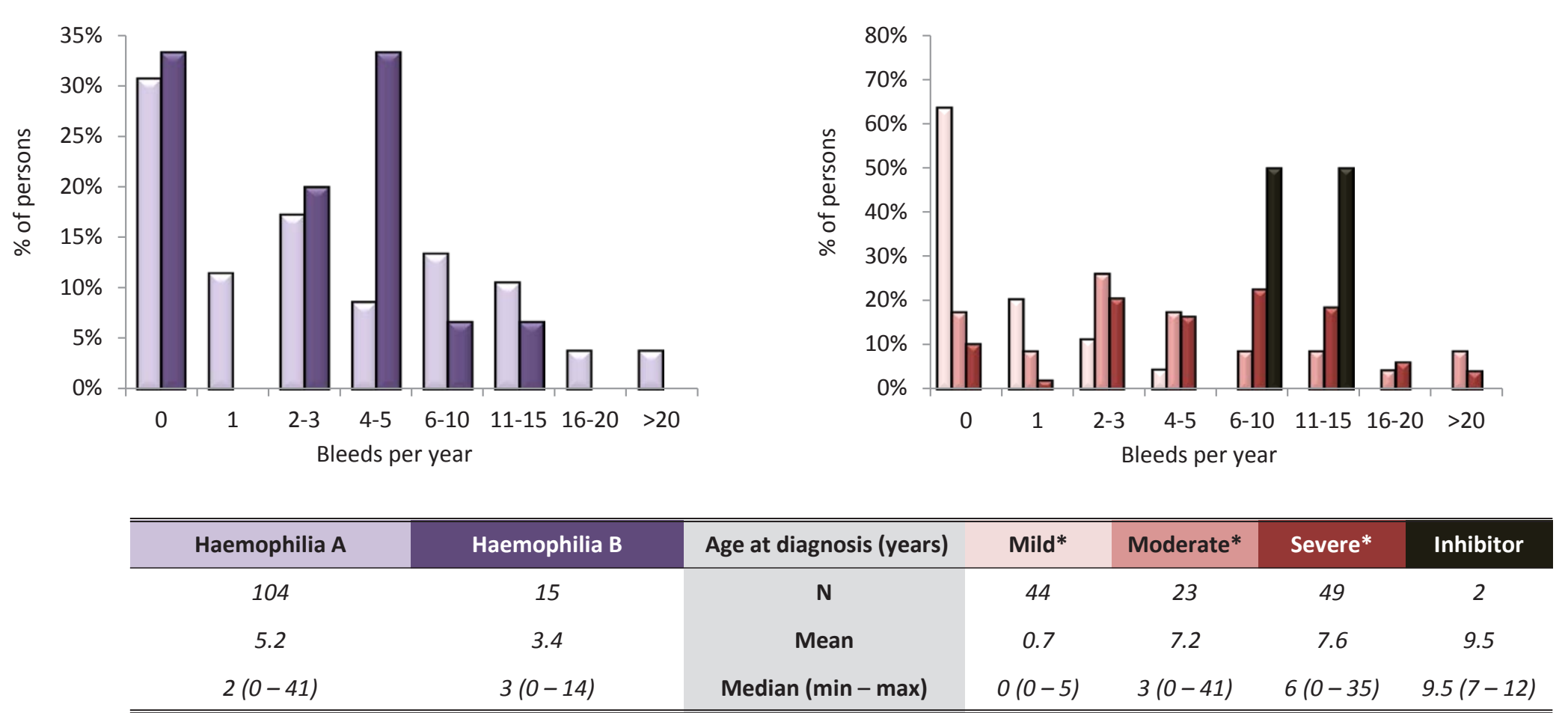
Thirteen per cent of Czech PWH are HCV positive (only two of them are children) and only 0.4 % of them are positive for HIV (all adults). Inhibitor rate is low in HA, with prevalence far below 5 % and incidence (excluding transient inhibitors) of 6 % in age group 0–18 years. Immune tolerance induction (ITI) treatment is the treatment of choice for Czech children with inhibitors with success rate of about 80 %. By-passing agents are available to treat bleedings in haemophiliacs with inhibitors.

Bleeding

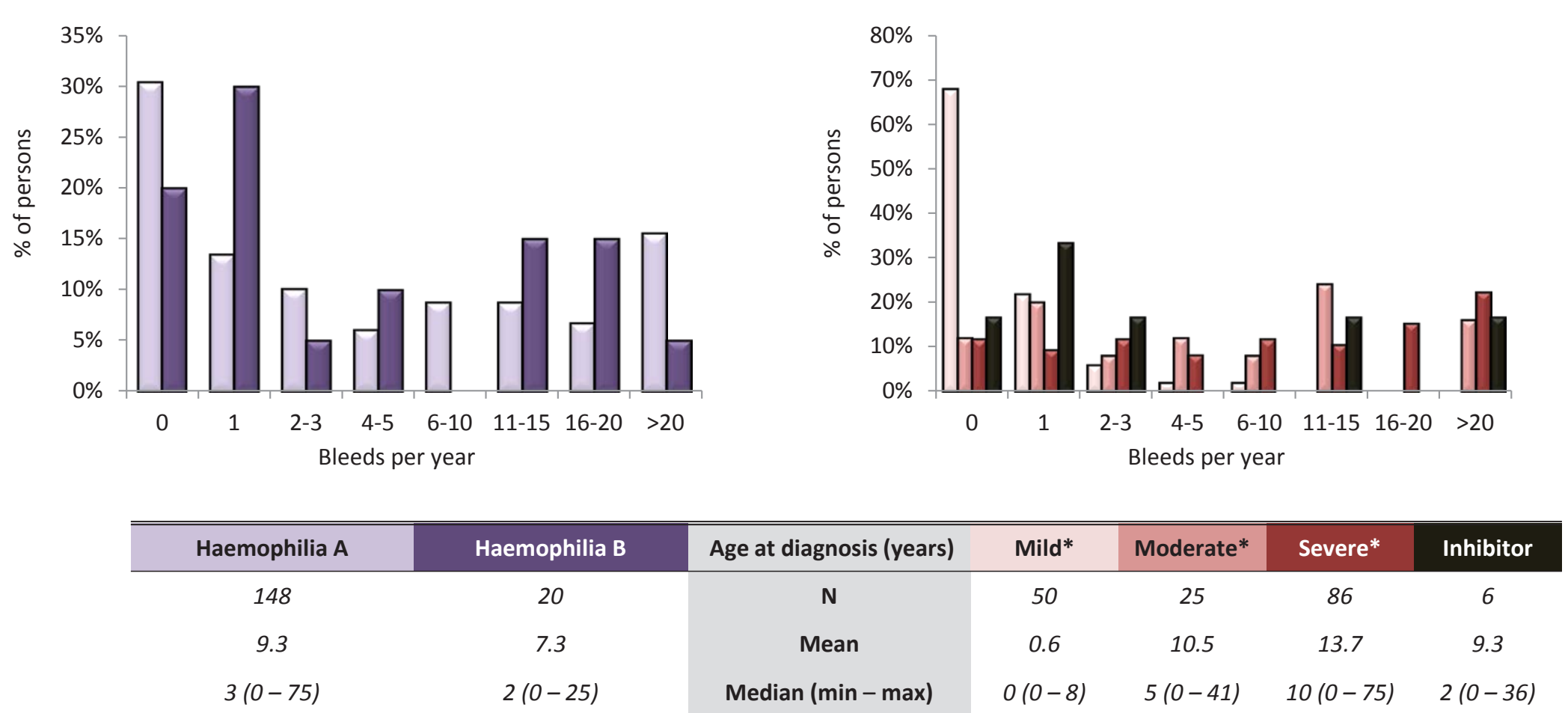
In 2011 Czech haemophiliacs bled in median 3 times per year (Haemophilia A: Adults: severe 10 /year, moderate 5/year, mild 0/year; Children: severe 6/year, moderate 3 /year, mild 0/year). (see Graphs 4, 5 and 6)



Graph 4. Bleeding frequency in all Czech PWH in 2011

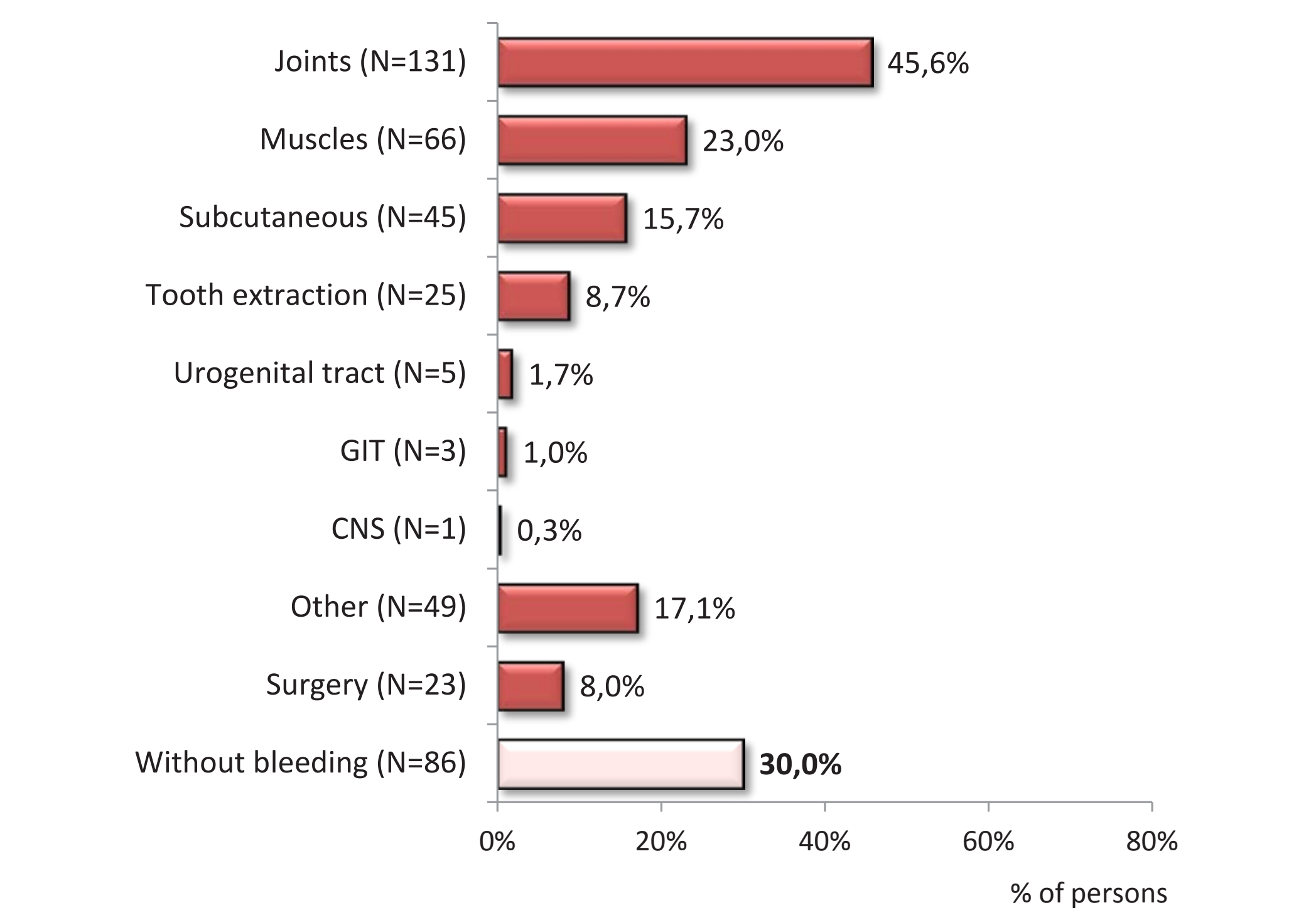


Graph 5. Bleeding frequency in Czech Children with Haemophilia in 2011



Graph 6. Bleeding in Czech Adult PWH in 2011

45.6 % of all episodes are joint bleeds. Last year no bleeding per year was recorded in 30.7 % of Czech PWH. (See graph 7)



Graph 7. Bleeding location in Czech PWH in 2011

Elective surgery and other interventions

Total hip and knee replacement as well as other surgeries are available for all, who need it, including patients with inhibitors.

References

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