Before 1990

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 became available also for Czech patients with haemophilia (PWH). In accord with WFH recommendations and later on with the European principles of haemophilia care the network of haemophilia centres (CSS; HFC) 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 of haemophilia, who needed it. Further step was the introduction of their 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 haemophilia care in the Czech Republic – 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 lost 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.

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 inaccessible also for blood born infections involving HUV
• 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 the haemophilia care population is shown on graphs 1 and 2.

Graph 2. Types 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 regimen.

Safety and inhibitors

Thirteen per cent of Czech PWH are HIV 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)

Conclusion

Authors want to thank all people involved in the Czech National Haemophilia Programme. Without their valuable work and efforts, no data would be available and we would not have any feedback of the haemophilia care in Czech Republic.

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.

References


Acknowledgements

This work was supported by the INTER DISCIPLINARY WORKING GROUP and INTER DISCIPLINARY WORKING GROUP – CZECH. The results shown in this publication reflect only the authors opinion and not necessarily the opinion of the INTER DISCIPLINARY WORKING GROUP and INTER DISCIPLINARY WORKING GROUP – CZECH.