## A. National Hemophilia Organization

| | |
|---|---|
| Organization name | Czech Society of Hemophilia |
| City | Prague |
| Country | Czech Republic |
| Phone | 420777078509 |
| E-mail | info@hemofilici.cz |
| This form completed by: | First name Martin<br>Last name Bohun<br>Email m.bohun@hemofilici.cz |

**Please** Click Here **to validate Organization contact information**

**The WFH would like to know how you collect the data you are providing for this survey. If you have a registry, we would like to know more about the registry.** A registry is a regularly updated centralized list of identified people with hemophilia (PWH) or inherited bleeding disorders. A registry includes information on personal details, diagnosis, treatment, and complications**.**

| | |
|---|---|
| What is the source of the numbers provided for this survey? | **Check one**<br>☐ Hemophilia Society and/or NMO registry or database<br>☒ Hospital(s)/HTC(s) registry or database<br>☐ Health Ministry registry or database<br>☐ Other (please describe): |
| How often is your database updated? | ☒ Ongoing update (can be updated anytime)<br>☐ Yearly update (the registry is updated once each year)<br>☐ Other (please describe): |
| Who updates the database? | ☐ Doctors update the database<br>☐ Patient organization updates the database<br>☒ Hospitals or clinics update the database<br>☐ Other (please describe): |

**Please** Click Here **to validate Data source**

## B. Identified patients

| **(Please DO NOT estimate or guess)** | **Number** | **Not known** |
|---|---|---|
| **1.** Total number of identified people with **hemophilia A or B,** or **type unknown** (PWH) | 1066 | ☐ |
| **2.** Number of identified people with **von Willebrand disease** (VWD) | 804 | ☐ |
| **3.** Number of identified people with other hereditary bleeding disorders (including rare factor deficiencies and inherited platelet disorders. See question 6 for the list of specific disorders.) | 77 | ☐ |
| Do you consider these numbers to be accurate? | Yes ☒ | Not sure ☐ |

**Please** Click Here **to validate number of patients**

**4. Number of people with Hemophilia and von Willebrand disease by age group**

| Age group | Number with hemophilia A | Number with hemophilia B | Number with hemophilia type unknown | Number with VWD |
|---|---|---|---|---|
| 0 - 4 years old | 41 | 7 | 0 | 10 |
| 5 - 13 years old | 102 | 19 | 0 | 65 |
| 14 - 18 years old | 57 | 8 | 0 | 40 |
| 19 - 44 years old | 432 | 51 | 0 | 371 |
| 45 years or older | 298 | 51 | 0 | 318 |
| Patients with age Unknown | 0 | 0 | 0 | 0 |
| No age data | ☐ | ☐ | ☐ | ☐ |

The age distribution of Hemophilia A, B and unknown should be equal to the number of PWH in question B1
The age distribution of vWD should be equal to the number of vWD in question B2

| | | |
|---|---|---|
| Do you consider these numbers to be accurate? | Yes ☐ | Not sure ☒ |
| **5.** Do you collect age data in a format that does not match question 4? (If you do collect age data in another format, please send it to the WFH in a separate attachment.) | | Yes ☐ |

**Please** Click Here **to validate Age section**

**6. Type of hereditary bleeding disorder**

The sum of *Male, Female,* and *Gender Unknown* should be equal to Total.

| Diagnosis | Total | Male | Female | Gender unknown | No data |
|---|---|---|---|---|---|
| Hemophilia A | 930 | 930 | 0 | 0 | ☐ |
| Hemophilia B | 136 | 136 | 0 | 0 | ☐ |
| Hemophilia, type unknown | 0 | 0 | 0 | 0 | ☐ |
| von Willebrand disease | 804 | 220 | 256 | 328 | ☐ |
| Factor I deficiency | 0 | 0 | 0 | 0 | ☐ |
| Factor II deficiency | 1 | 0 | 1 | 0 | ☐ |
| Factor V deficiency | 5 | 1 | 4 | 0 | ☐ |
| Factor V+VIII deficiency | 0 | 0 | 0 | 0 | ☐ |
| Factor VII deficiency | 33 | 14 | 19 | 0 | ☐ |
| Factor X deficiency | 4 | 3 | 1 | 0 | ☐ |
| Factor XI deficiency | 17 | 9 | 8 | 0 | ☐ |
| Factor XIII deficiency | 2 | 1 | 1 | 0 | ☐ |
| Rare factor deficiency: type unknown | 15 | 4 | 11 | 0 | ☐ |
| Platelet disorders: Glanzmann's thrombasthenia | | | | | ☐ |
| Platelet disorders: Bernard Soulier Syndrome | | | | | ☐ |
| Platelet disorders: other or unknown | | | | | ☐ |

The sum of Totals Hemophilia A, B, and type unknown should be equal to the number of PWH in question B1.
The Total of vWD should be equal to the number of vWD in question B2
The sum of Total of the all other bleeding and platelets disorders should be equal to the number of OBD in question B3

| | | |
|---|---|---|
| Do you consider these numbers to be accurate? | Yes ☒ | Not sure ☐ |

**Please** Click Here **to validate Gender section**

**7. How are patients with rare bleeding disorders (deficiency in FI, FII, FV, FV+VIII, FVII, FX, FXI FXIII) classified?**

| | | | |
|---|---|---|---|
| Factor level measurements ☒ | Clinical diagnosis ☐ (bleeding, family history) | Other ☐ (please describe): | No data ☐ |

**How are patients with von Willebrand Disease classified?**

| | | | |
|---|---|---|---|
| Factor level measurements ☒ | Severe bleeding symptoms ☐ | Other ☐ (please describe): | No data ☐ |

**8. Number of identified people with hemophilia by diagnosis of severity**

There are three levels of **severity** of hemophilia: **mild, moderate,** and **severe**. The severity of hemophilia depends on the amount of clotting factor in the person's blood.

A person (male or female) with 5-40 per cent of the normal amount of clotting factor has **mild** hemophilia.

A person (male or female) with between 1-5 per cent of the normal amount of clotting factor has **moderate** hemophilia.

A person (male or female) with less than 1 per cent of the normal amount of clotting factor has **severe** hemophilia.

A woman who has less than 40 per cent of the normal level of clotting factor is no different from a man with the same factor levels—she has hemophilia.

| Type of hemophilia | Mild (factor level above 5%) | Moderate (factor level 1% to 5%) | Severe (factor level below 1%) | Severity unknown | No Data |
|---|---|---|---|---|---|
| Hemophilia A male | 438 | 148 | 308 | 36 | ☐ |
| Hemophilia A female | 0 | 0 | 0 | 0 | ☐ |
| Hemophilia B male | 36 | 44 | 50 | 6 | ☐ |
| Hemophilia B female | 0 | 0 | 0 | 0 | ☐ |

The sum of Hemophilia A Male mild, moderate, severe and unknown should be equal to number of Hemophilia A Male in question 6
The sum of Hemophilia A Female mild, moderate, severe and unknown should be equal to number of Hemophilia A female in question 6
The sum of Hemophilia B Male mild, moderate, severe and unknown should be equal to number of Hemophilia B Male in question 6
The sum of Hemophilia B Female mild, moderate, severe and unknown should be equal to number of Hemophilia B female in question 6

| | | |
|---|---|---|
| Do you consider these numbers to be accurate? | Yes ☒ | Not sure ☐ |

**9. Number of severe VWD patients**

| Total number of severe (type 3) VWD patients | Number of VWD patients receiving replacement therapy | Number of VWD patients with severe bleeding symptoms | No Data |
|---|---|---|---|
| **23** | 138 | 71 | ☐ |

| | | |
|---|---|---|
| Do you consider these numbers to be accurate? | Yes ☐ | Not sure ☒ |

**10. INHIBITORS: Number of identified people with hemophilia with current clinically significant inhibitors. (Patients who do not respond to normal treatment.)**

| Type of hemophilia | Total number with active inhibitors | New cases of inhibitors in 2014 | No Data |
|---|---|---|---|
| Hemophilia A | 16 | 3 | ☐ |
| Hemophilia B | 2 | 0 | ☐ |

**Please** Click Here **to validate classification, severity and inhibitors**

**11. Products used to treat hemophilia: How many patients were treated with the following products? (Please note: we are asking for a number, not a percentage.)**

| Treatment product | Number treated | Product is available | Product is used | Product is not used |
|---|---|---|---|---|
| Plasma | | ☐ | ☐ | ☒ |
| Cryoprecipitate | | ☐ | ☐ | ☒ |
| Plasma-derived concentrate | 438 | ☒ | ☒ | ☐ |
| Recombinant concentrate | 219 | ☒ | ☒ | ☐ |
| DDAVP (Desmopressin) | | ☐ | ☐ | ☐ |

**12. Products used to treat VWD: How many patients were treated with the following products? (Please note: we are asking for a number, not a percentage.)**

| Treatment product | Number treated | Product is available | Product is used | Product is not used |
|---|---|---|---|---|
| Plasma | | ☐ | ☐ | ☒ |
| Cryoprecipitate | | ☐ | ☐ | ☒ |
| Plasma-derived concentrate | 138 | ☒ | ☒ | ☐ |
| DDAVP (Desmopressin) | | ☐ | ☐ | ☐ |

**13. HIV and hepatitis C infection among living people with hemophilia (Please note: we are asking for a number, not a percentage.)**

| Infectious Disease | Number of people infected | Number of people tested | No Data |
|---|---|---|---|
| HIV | 3 | 823 | ☐ |
| Hepatitis C | 205 | 1010 | ☐ |

**14. HIV and hepatitis C infection among living people with von Willebrand disease (Please note: we are asking for a number, not a percentage.)**

| Infectious Disease | Number of people infected | Number of people tested | No Data |
|---|---|---|---|
| HIV | 0 | 230 | ☐ |
| Hepatitis C | 2 | 399 | ☐ |

**15. Number and cause of deaths of people with bleeding disorders (January 1-December 31, 2014)**

| Cause of death | Number of people with Hemophilia A & B | Number of people with von Willebrand disease | Number of people with other inherited bleeding disorders |
|---|---|---|---|
| Bleeding | | | |
| HIV | | | |
| Liver disease | | | |
| Other causes | 2 | 0 | 0 |

**Please** Click Here **to validate products, HIV, HCV, and cause of death sections**

## C. Hemophilia Care System in Your Country

A Hemophilia Treatment Centre (HTC) is a medical centre providing basic diagnosis and treatment for inherited bleeding disorders.

A Hemophilia Comprehensive Care Centre (HCCC) is a medical centre providing a full range of facilities for the diagnosis and management of inherited bleeding disorders.

| | |
|---|---|
| **16**. How many **hemophilia treatment centres** are there in your country? | 9 |
| Of these, how many are **hemophilia comprehensive care centres**? | 3 |
| Percentage of hemophilia patients with access to hemophilia treatment centres: | 100 |

**Prophylaxis** is regular, long-term treatment with clotting factor concentrates to prevent bleeds. Please indicate if the percentage provided is precise or an estimate.

| | | | |
|---|---|---|---|
| **17**. What percentage of eligible children (under age 18) **with severe hemophilia** are on prophylaxis? | 84 | Precise: ☒ Estimate: ☐ | Not known ☐ |
| What percentage of eligible adults (over age 18) **with severe hemophilia** are on prophylaxis? | 47 | Precise: ☒ Estimate: ☐ | Not known ☐ |

**Please** Click Here **to validate Care section**

## D. The Cost and Use of Factor Concentrates

| **18. Annual usage of factor concentrates** | **Factor VIII** | **Not known** | **Factor IX** | **Not known** |
|---|---|---|---|---|
| **IN TOTAL** how many international units (IU) of factor concentrates were used in your country in 2014? | 51 721 750 | ☐ | 5 415 520 | ☐ |
| How many international units of **plasma-derived** concentrates were used in your country in 2014? | 33 325 500 | ☐ | 4 814 700 | ☐ |
| How many international units of **recombinant** concentrates were used in your country in 2014? | 18 396 250 | ☐ | 600 820 | ☐ |

The sum of Total of FVIII should be equal to sum of FVIII plasma-derived and FVIII recombinant
The sum of Total of FIX should be equal to sum of FIX plasma-derived and FIX recombinant

| | | | | |
|---|---|---|---|---|
| Of the number reported above how many international units were **humanitarian aid**? | 0 | ☐ | 0 | ☐ |

| | | |
|---|---|---|
| Do you consider these numbers to be accurate? | Yes ☒ | Not sure ☐ |

**PLEASE NOTE:** If a product used in your country is not listed, please add it at the bottom of the appropriate table.

| | | |
|---|---|---|
| Currency: CZK | Tax included? No ☐ Yes ☐ | Tax rate: |

**Please** Click Here **to validate Factors section**

**19. Factor VIII Concentrates used in 2014**

(Please check the box on the left if a product is used, and if known, fill out the cost per international unit in the currency used to purchase the product. Please indicate if this price includes tax.)

| Used | Brand Name | Manufacturer | Price per IU |
|---|---|---|---|
| ☐ | Aafact | Sanquin | |
| ☒ | Advate rAHF PFM | Baxter Bioscience | |
| ☐ | Aleviate | CSL Behring | |
| ☐ | Alphanate | Grifols | |
| ☐ | Amofil | Sanquin OY | |
| ☐ | Beriate P | CSL Behring | |
| ☐ | BIOSTATE | CSL Bioplasma | |
| ☐ | Conco-eight-HT | Benesis | |
| ☐ | Confact F | Kaketsuken | |
| ☐ | Cross Eight M | Japanese Red Cross | |
| ☐ | Elocta/Eloctate | Biogen Idec | |
| ☐ | Emoclot D.I. | Kedrion | |
| ☐ | FACTANE | LFB | |
| ☐ | Factor 8 Y | BioProducts Lab. | |
| ☐ | Faktor VIII SDH Intersero | Intersero | |
| ☒ | Fanhdi | Grifols | |
| ☐ | GreenEight | GreenCross | |
| ☐ | GreenGene | GreenCross | |
| ☐ | GreenMono | Greencross Corp | |
| ☒ | Haemate P (= Haemate HS) | CSL Behring | |
| ☒ | Haemoctin SDH | Biotest | |
| ☐ | Haemosolvate Factor VIII | National Bioproducts | |
| ☒ | Helixate NexGen = Helixate FS | CSL Behring | |
| ☐ | Hemofil M AHF | Baxter BioScience | |
| ☐ | HEMORAAS SD plus H | Shanghai RAAS | |
| ☐ | HEMORAAS-HP, SD plus H | Shanghai RAAS | |
| ☐ | HEMORAAS-IP, SD plus H | Shanghai RAAS | |
| ☐ | Humate P | CSL Behring | |
| ☐ | Humafaktor 8 | Human BioPlazma | |
| ☐ | Human Coagulation Factor VIII | Baltijas Terapeitiskais Serviss | |
| ☒ | Immunate | Baxter BioScience | |
| ☐ | Koate DVI | Talecris | |
| ☒ | Kogenate FS = KOGENATE Bayer (in EU) | Bayer | |
| ☐ | Monoclate P | CSL Behring | |
| ☐ | Novoeight | NovoNordisk | |
| ☒ | Octanate | Octapharma | |
| ☐ | Octanativ-M | Octapharma | |

| | | | |
|---|---|---|---|
| ☐ | Octavi SD | Octapharma | |
| ☐ | Optivate | Bio Products Laboratory | |
| ☒ | Recombinate rAHF | Baxter BioScience | |
| ☐ | ReFacto AF | Pfizer (Wyeth) | |
| ☐ | Replenate | Bio Products Laboratory | |
| ☒ | Wilate | Octapharma | |
| ☐ | Xyntha | Pfizer (Wyeth) | |
| ☐ | Other: | | |

**20. Factor IX Concentrates used in 2014**

(Please check the box on the left if a product is used, and if known, fill out the cost per international unit in your currency.)

| Used | Brand Name | Manufacturer | Price per IU |
|---|---|---|---|
| ☐ | Aimafix | Kedrion | |
| ☐ | AlphaNine SD | Grifols | |
| ☐ | Alprolix | Biogen Idec | |
| ☐ | BeneFIX | Wyeth | |
| ☐ | Berinin-P = Berinin HS | CSL Behring | |
| ☐ | BETAFACT | LFB | |
| ☐ | Christmassin-M | Benesis | |
| ☐ | Factor IX Grifols | Grifols | |
| ☐ | Faktor IX SDN | Biotest | |
| ☐ | Fixnove | Baxter | |
| ☐ | Hemo-B-RAAS | Shanghai RAAS | |
| ☐ | Haemonine | Biotest | |
| ☒ | Immunine | Baxter BioScience | |
| ☐ | MonoFIX-VF | CSL Bioplasma | |
| ☒ | Mononine | CSL Behring | |
| ☐ | Nanofix | Octapharma | |
| ☐ | Nanotiv | Octapharma | |
| ☐ | Nonafact | Sanquin | |
| ☐ | Novact M | Kaketsuken | |
| ☐ | Octafix | Octapharma | |
| ☒ | Octanine F | Octapharma | |
| ☐ | Replenine – VF | BioProducts Lab. | |
| ☐ | Other: | | |

**21. Prothrombin Complex Concentrates used in 2014**

(Please check the box on the left if a product is used, and if known, fill out the cost per international unit in your currency.)

| Used | Brand Name | Manufacturer | Price per IU |
|---|---|---|---|
| ☐ | Bebulin VH | Baxter BioScience | |
| ☐ | Beriplex P/N | CSL Behring | |
| ☐ | Cofact | Sanquin | |
| ☐ | Facnyne | Greencross Corp | |
| ☐ | Haemosolvex Factor IX | National Bioproducts | |
| ☐ | HT DEFIX | SNBTS | |
| ☐ | Kanokad Confidex | LFB | |
| ☐ | KASKADIL | LFB | |
| ☐ | Octaplex | Octapharma | |
| ☐ | PPSB-HT | Nihon Pharmaceutical | |
| ☐ | PPSB-human SD/Nano 300/600 | German Red Cross NSTOB | |
| ☐ | Profilnine SD | Grifols | |
| ☐ | Proplex – T | Baxter BioScience | |
| ☐ | Prothrombinex PXT | CSL Bioplasma | |
| ☐ | Prothrombinex- VF | CSL Bioplasma | |
| ☐ | Prothromplex-T | Baxter BioScience | |
| ☐ | Prothroraas | Shanghai RAAS | |
| ☐ | UMAN Complex D.I. | Kedrion | |
| ☐ | Other: | | |

**22. Other Products used in 2014**

(Please check the box on the left if a product is used, and if known, fill out the cost per international unit in your currency.)

| Used | Brand Name | Manufacturer | Price per IU |
|---|---|---|---|
| ☐ | Aryoseven | Aryogen | |
| ☐ | Clottafact Wilstart | LFB | |
| ☐ | Clottagen (fibrinogen) | LFB | |
| ☐ | Coagil 7 (activated factor VII) | Pharmstandard | Price per vial:<br>Vial size: |
| ☐ | FACTEUR VII | LFB | |
| ☐ | Factor VII | Baxter BioScience | |
| ☐ | Factor VII | Bio Products | |
| ☐ | Factor X P Behring | CSL Behring | |
| ☐ | Factor XI | Bio Products | |
| ☒ | FEIBA | Baxter | |
| ☐ | Fibrinogen HT | Benesis | |
| ☐ | Fibrogammin P (=Fibrogammin HS) (Factor XIII) | CSL Behring | |
| ☐ | FIBRORAAS (fibrinogen) | Shanghai RAAS | |
| ☐ | Haemocomplettan P = Haemocomplettan HS (fibrinogen) | CSL Behring | |
| ☐ | HEMOLEVEN (Factor XI) | LFB | |

| | | | |
|---|---|---|---|
| ☒ | NovoSeven (=Niastase) (activated factor VII) | NovoNordisk | Price per vial:<br>Vial size: |
| ☐ | Riastap | CSL Behring | |
| ☐ | Tretten rXIII | NovoNordisk | |
| ☐ | WILFACTIN (Von Willebrand Factor) | LFB | |
| ☐ | Other: | | |

**Please return to:** globalsurvey@wfh.org
**Fax:** (514-875-8916)
or return by mail to: **World Federation of Hemophilia**
1425 René Lévesque Boulevard West, suite 1010,
Montréal, Québec, H3G 1T7, Canada

## Glossary of terms

**Bernard-Soulier syndrome:** A severe congenital bleeding disorder characterized by thrombocytopenia and large platelets, due to a defect in the platelet glycoprotein 1b/V/IX receptor.

**Cryoprecipitate:** A fraction of human blood prepared from fresh plasma. Cryoprecipitate is rich in factor VIII, von Willebrand factor, and fibrinogen (factor I). It does not contain factor IX.

**Desmopressin (DDAVP):** A synthetic hormone used to treat most mild cases of von Willebrand disease and mild hemophilia A. It is administered intravenously or by subcutaneous injection or by intranasal spray.

**Factor concentrates:** These are fractionated, freeze-dried preparations of individual clotting factors or groups of factors derived from donated blood.

**Glanzmann's thrombasthenia:** A severe congenital bleeding disorder in which the platelets lack glycoprotein IIb/IIIa, the blood platelet count is normal, but their function is very abnormal.

**Hemophilia A:** A condition resulting from factor VIII deficiency, also known as classical hemophilia.

**Hemophilia B:** A condition resulting from factor IX deficiency, also known as Christmas disease.

**Hemophilia treatment centre:** A specialized medical centre that provides diagnosis, treatment, and care for people with hemophilia and other inherited bleeding disorders.

**HIV:** Human immunodeficiency virus. The virus that causes AIDS.

**Identified person:** A living person known to have hemophilia, von Willebrand disease, or another bleeding disorder.

**Inhibitors:** A PWH has inhibitors when their body's immune system attacks the molecules in factor concentrate, rendering it ineffective.

**International Unit (IU)**: A standardized measurement of the amount of factor VIII or IX contained in a vial. Usually marked on vials as 250 IU, 500 IU, 1000 IU or 2000 IU.

**Mild hemophilia:** Condition resulting from a level of factor VIII or factor IX clotting activity below normal but above 5% of normal activity in the bloodstream. (National definitions differ on the upper limit for mild hemophilia, ranging from 24% to 50%. The normal range of factor VIII or IX is 50 to 200%)

**Moderate hemophilia:** Condition resulting from a level of factor VIII or factor IX clotting activity between 1 to 5 % of normal activity in the bloodstream.

**Plasma-derived products:** Factor concentrates that contain factor VIII or IX that have been fractionated from human blood.

**PWH:** Person with hemophilia

**Recombinant products:** Factor concentrates that contain factor VIII or IX that have been artificially produced and are, therefore, not derived from human blood.

**Registry:** A database or record of identified people with hemophilia or inherited bleeding disorders. A registry includes information on personal details, diagnosis, treatment and complications.

**Severe hemophilia:** Condition resulting from a level of factor VIII or factor IX clotting activity of less than 1 % in the bloodstream.

**von Willebrand disease (VWD):** An inherited bleeding disorder resulting from a defect or deficiency of von Willebrand factor.