WFH
WORLD FEDERATION OF HEMOPHILIA
FÉDÉRATION MONDIALE DE L'HÉMOPHILIE
FEDERACIÓN MUNDIAL DE HEMOFILIA

# Annual Global Survey 2016

Please return to WFH by May 30, 2017

## A. National Hemophilia Organization

| | |
|---|---|
| Organization name | Czech Society of Hemophilia |
| City | Prague |
| Country | Czechia |
| Phone | +420603580980 |
| E-mail | info@hemofilici.cz |
| This form completed by: | First name Vladimír<br>Last name Dolejš<br>Email vladimir@dolejs.org |

**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): |
| Have all the identified patients in your country been included in this report? If not, please explain. | Yes ☒ No ☐<br>Please explain: |

**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) | 1076 | ☐ |
| **2.** Number of identified people with **von Willebrand disease** (VWD) | 818 | ☐ |
| **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.) | 109 | ☐ |
| 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 | 45 | 9 | 0 | 8 |
| 5 - 13 years old | 107 | 17 | 0 | 63 |
| 14 - 18 years old | 61 | 9 | 0 | 45 |
| 19 - 44 years old | 420 | 48 | 0 | 377 |
| 45 years or older | 304 | 56 | 0 | 325 |
| 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 | 937 | 937 | 0 | 0 | ☐ |
| Hemophilia B | 139 | 139 | 0 | 0 | ☐ |
| Hemophilia, type unknown | 0 | 0 | 0 | 0 | ☐ |
| von Willebrand disease | 818 | 252 | 313 | 253 | ☐ |
| Factor I deficiency | | | | | ☒ |
| Factor II deficiency | 1 | 0 | 1 | 0 | ☐ |
| Factor V deficiency | 8 | 1 | 7 | 0 | ☐ |
| Factor V+VIII deficiency | 0 | | | | ☐ |
| Factor VII deficiency | 52 | 21 | 31 | 0 | ☐ |
| Factor X deficiency | 2 | 1 | 1 | 0 | ☐ |
| Factor XI deficiency | 19 | 9 | 10 | 0 | ☐ |
| Factor XIII deficiency | 2 | 1 | 1 | 0 | ☐ |
| Rare factor deficiency: type unknown | 25 | 7 | 18 | 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
A woman who has less than 40 percent of the normal level of clotting factor would be considered a person with hemophilia. A woman with more than 40% FVIII is considered a carrier and should not be included in this report.

| | | |
|---|---|---|
| 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 gender and 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 percent of the normal level of clotting factor would be considered a person with hemophilia. A woman with more than 40% FVIII is considered a carrier and should not be included in this report.

| 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 | 331 | 70 | 248 | 288 | ☐ |
| Hemophilia A female | 0 | 0 | 0 | 0 | ☐ |
| Hemophilia B male | 30 | 30 | 38 | 41 | ☐ |
| 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 |
|---|---|---|---|
| **20** | 127 | 35 | ☐ |

| | | |
|---|---|---|
| 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 2016 | No Data |
|---|---|---|---|
| Hemophilia A | 21 | 5 | ☐ |
| Hemophilia B | 2 | 0 | ☐ |

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

**11. Availability and usage of products to treat hemophilia**

| Treatment product | Product is available | Product is used | Number of patients treated with product indicated | No data |
|---|---|---|---|---|
| Plasma | ☒ | ☐ | | ☐ |
| Cryoprecipitate | ☒ | ☐ | | ☐ |
| Plasma-derived concentrate | ☒ | ☒ | 375 | ☐ |
| Recombinant concentrate (excluding prolonged half-life) | ☒ | ☒ | 214 | ☐ |
| Recombinant concentrate (prolonged half-life) | ☒ | ☒ | 2 | ☐ |
| DDAVP (Desmopressin) | ☒ | ☒ | | ☒ |

**PLEASE NOTE:** We are asking for the number of patients treated, not a percentage. Please provide your best estimate.

**12. Availability and usage of products to treat VWD**

| Treatment product | Product is available | Product is used | Number of patients treated with product indicated | No data |
|---|---|---|---|---|
| Plasma | ☒ | ☐ | | ☐ |
| Cryoprecipitate | ☒ | ☐ | | ☐ |
| Plasma-derived concentrate | ☒ | ☒ | 127 | ☐ |
| DDAVP (Desmopressin) | ☒ | ☒ | | ☒ |

**PLEASE NOTE:** We are asking for the number of patients treated, not a percentage. Please provide your best estimate.

**13. HIV infection**

| | Hemophilia A or B, or type unknown | von Willebrand disease | Other hereditary bleeding disorders |
|---|---|---|---|
| Total number of people living with HIV | 3 | 0 | 0 |
| New HIV infections in 2016 | 0 | 0 | 0 |

**14. Hepatitis C infection**

| | Hemophilia A or B, or type unknown | von Willebrand disease | Other hereditary bleeding disorders |
|---|---|---|---|
| Total number of people infected with hepatitis C[1] | 210 | 2 | 0 |
| Total number of people with currently active hepatitis C[2] | 60 | 1 | 0 |
| New hepatitis C infections in 2016 | 0 | 0 | 0 |

[1]Hepatitis C antibody positive at any time
[2]Still PCR positive: patients who have not cleared the virus spontaneously or after treatment

**15. Number and cause of deaths of people with bleeding disorders (January 1-December 31, 2016)**

| 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 | 0 | 0 | 0 |
| HIV | 0 | 0 | 0 |
| Liver disease | 0 | 0 | 0 |
| Other causes | 3 | 0 | 0 |

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

## C. Hemophilia Care System in Your Country

We define as Hemophilia Treatment Centre (HTC) a medical centre providing any level of care (including basic diagnosis and treatment) for inherited bleeding disorders. Please provide the number of all such centres in your country. Please also indicate how many of those centers have **direct access, within the same structure,** to at least the following: hemophilia doctor, nurse, physiotherapist, social worker, and special coagulation laboratory.

| | |
|---|---|
| **16**. How many **hemophilia treatment centres** are there in total in your country? | 11 |
| How many of the **hemophilia treatment centres** you have indicated above have **direct access, within the same structure,** to a hemophilia doctor, nurse, physiotherapist, social worker, and special coagulation laboratory? | 11 |
| Which percentage of the hemophilia patients in your country has access to a hemophilia treatment centre: | 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 children (under age 18) **with severe hemophilia** are on prophylaxis? | 86 | Precise: ☒ Estimate: ☐ | Not known ☐ |
| What percentage of adults (over age 18), **with severe hemophilia** are on prophylaxis? | 57 | Precise: ☒ Estimate: ☐ | Not known ☐ |
| What is the most common dose (IU/kg) of factor administered and frequency? | Prophy adults:18 IU/kg 3/week, kids 25 IU/kg 3/week | | |

**Immune tolerance induction (ITI)** is the administration of FVIII or FIX concentrate in patients with inhibitors to eradicate the inhibitors. Please indicate the total percentage of patients with inhibitors receiving ITI in your country and the number of patients having received ITI during last year and indicate if what you provided is precise or an estimate.

| | | | |
|---|---|---|---|
| **18**. What **percentage** of patients **with inhibitors** are receiving or have ever received immune tolerance induction? | 70 | Precise: ☐ Estimate: ☒ | Not known ☐ |
| How many patients **with inhibitors** have received immune tolerance induction in the last year? | 6 | Precise: ☒ Estimate: ☐ | Not known ☐ |

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

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## D. The Cost and Use of Factor Concentrates

Okomentoval(a): [Factor1]: Please indicate if tax is included

| 19 A. Annual usage of purchased factor concentrates (please do not include donated factor) | Factor VIII | Not known | Factor IX | Not known |
|---|---|---|---|---|
| **IN TOTAL** how many international units (IU) of factor concentrates were used in your country in 2016 (excluding donated factor)? | 59791435 | ☐ | 6617556 | ☐ |
| How many international units of **plasma-derived** concentrates were used in your country in 2016 (excluding donated factor)? | 27827829 | ☐ | 6027050 | ☐ |
| How many international units of **recombinant** concentrates were used in your country in 2016 (excluding donated factor)? | 31963606 | ☐ | 590506 | ☐ |

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

| 19 B. Annual usage of donated factor concentrates | Factor VIII | Not known | Factor IX | Not known |
|---|---|---|---|---|
| How many international units of **donated factor** concentrates (plasma-derived or recombinant) from all sources, including **Humanitarian Aid,** were used in your country in 2016? | 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**

**20. Factor VIII Concentrates used in 2016**

(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 | Baxalta (Baxter Bioscience) | |
| ☐ | Adynovate | Baxalta (Baxter Bioscience) | |
| ☐ | Aleviate | CSL Behring | |
| ☐ | Alphanate | Grifols | |
| ☐ | Amofil | Sanquin OY | |
| ☐ | Bioclot A | Biofarma | |
| ☐ | 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 | |
| ☐ | HEMO-8R | HEMOBRAS | |
| ☐ | Hemofil M AHF | Baxalta (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 | Baxalta (Baxter Bioscience) | |
| ☐ | Koate DVI | Talecris | |

| | | | |
|---|---|---|---|
| ☒ | Kogenate FS = KOGENATE Bayer (in EU) | Bayer | |
| ☐ | Monoclate P | CSL Behring | |
| ☐ | Novoeight | NovoNordisk | |
| ☒ | Nuwiq | Octapharma | |
| ☒ | Octanate | Octapharma | |
| ☐ | Octanativ-M | Octapharma | |
| ☐ | Octavi SD | Octapharma | |
| ☐ | Octofactor | Generium/Pharmstandart | |
| ☐ | Optivate | Bio Products Laboratory | |
| ☐ | FVIII by Quimbiotec | Quimbiotec | |
| ☐ | Recombinate rAHF | Baxalta (Baxter Bioscience) | |
| ☒ | ReFacto AF | Pfizer (Wyeth) | |
| ☐ | Replenate | Bio Products Laboratory | |
| ☐ | TBSF purity factor, Koate DVI | Grifols | |
| ☐ | UNC Hemoderivados | Laboratorio de Hemoderivados de Universidad Nacional de Córdoba | |
| ☐ | Voncento | CSL Behring | |
| ☐ | Western Province factor8 VIAHF | Western Province Blood transfusion Service | |
| ☒ | Wilate | Octapharma | |
| ☐ | Xyntha | Pfizer (Wyeth) | |
| ☐ | Other: | | |

**PLEASE NOTE:** For "Other", please provide the Brand Name and Manufacturer.

**21. Factor IX Concentrates used in 2016**

(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 | |
| ☐ | Clotnine | Hemarus | |
| ☐ | Factor IX Grifols | Grifols | |
| ☐ | Faktor IX SDN | Biotest | |
| ☐ | Fixnove | Baxalta (Baxter Bioscience) | |
| ☐ | Hemo-B-RAAS | Shanghai RAAS | |
| ☐ | Haemonine | Biotest | |
| ☐ | Humafactor IX | Kedrion | |
| ☒ | Immunine | Baxalta (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. | |
| ☒ | Rixubis | Baxalta (Baxter Bioscience) | |
| ☐ | Other: | | |

**PLEASE NOTE:** For "Other", please provide the Brand Name and Manufacturer.

**22. Prothrombin Complex Concentrates used in 2016**

(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 | Baxalta (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 | Baxalta (Baxter Bioscience) | |
| ☐ | Prothrombinex PXT | CSL Bioplasma | |
| ☐ | Prothrombinex- VF | CSL Bioplasma | |
| ☒ | Prothromplex-T | Baxalta (Baxter Bioscience) | |
| ☐ | Prothroraas | Shanghai RAAS | |
| ☐ | UMAN Complex D.I. | Kedrion | |
| ☐ | Other: | | |

**PLEASE NOTE:** For "Other", please provide the Brand Name and Manufacturer.

**23. Other Products used in 2016**

(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 | |

| | | | |
|---|---|---|---|
| ☐ | Byclot (1.5mg) | Kaketusken | |
| ☒ | Ceprotin | Baxalta (Baxter Bioscience) | |
| ☐ | Clottafact Wilstart | LFB | |
| ☐ | Clottagen (fibrinogen) | LFB | |
| ☐ | Coagil 7 (activated factor VII) | Pharmstandard | Price per vial:<br>Vial size: |
| ☐ | FACTEUR VII | LFB | |
| ☒ | Factor VII | Baxalta (Baxter Bioscience) | |
| ☐ | Factor VII | Bio Products | |
| ☒ | Factor X P Behring | CSL Behring | |
| ☐ | Factor XI | Bio Products | |
| ☒ | FEIBA | Baxalta (Baxter Bioscience) | |
| ☐ | 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 | |
| ☒ | Kovaltry | Bayer | |
| ☒ | 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 NOTE:** For "Other", please provide the Brand Name and Manufacturer.

**Please return to:**

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Canada

**Please provide your feedback on the WFH Annual Global Survey data collection system.**

Comments:

| |
|---|
| |

## 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.