A. National Hemophilia Organization

Organization name	Czech Society of Hemophilia
Address	U Nemocnice 1 (UHKT)
City	Praha 2
State, Province, Region, Prefecture, County	
Postal/ZIP Code	CZ-128 20
Country	Czech Republic
Phone	+420603580980
Fax	
E-mail	info@hemofilici.cz
Website	www.hemofilici.cz

B. Identified patients

(Please DO NOT estimate or guess)	**Number**	**Not known**
1. Number of identified people with **hemophilia A** and **B** (PWH)	1031	☐
2. Number of identified people with **von Willebrand disease** (vWD)	478	☐
3. Number of identified people with other hereditary bleeding disorders (including rare factor deficiencies and inherited platelet disorders)	49	☐
Do you consider these numbers to be accurate?	Yes ☒	Not sure ☐

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**.**

4. What is the source of the numbers provided for this survey?	**Check one** ☐ A **registry** of all PWH and other inherited bleeding disorders in your country. ☒ A **registry** of all PWH and other inherited bleeding disorders in your country's hemophilia treatment centres. ☐ Count information provided by all of your country's hemophilia treatment centres ☐ Count information provided by some of your country's hemophilia treatment centres. ☐ Other (Describe):
Is your database updated throughout the year or only once per year?	☐ Ongoing update (can be updated anytime) ☒ Yearly update (the registry is updated once each year) ☐ Other (please describe):
Who updates the database?	☒ Doctors update the database ☐ Patient organization updates the database ☐ Hospitals or clinics update the database ☐ Other (please describe):

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

Age group	Number with hemophilia A	Number with hemophilia B	Number with VWD
0-4 years old	41	7	5
5 - 13 years old	101	13	35
14 - 18 years old	62	9	17
19 - 44 years old	384	55	233
45 years or older	310	49	188
Patients with age unknown	0	0	0
No age data	☐	☐	☐

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

6.Type of hereditary bleeding disorder

Diagnosis	Total	Male	Female	Gender unknown	No data
Hemophilia A	898	898	0		☐
Hemophilia B	133	133	0		☐
Hemophilia, type unknown	0				☐
von Willebrand disease	478	212	266		☐
Factor I deficiency					☒
Factor II deficiency					☒
Factor V deficiency					☒
Factor V+VIII deficiency					☒
Factor VII deficiency					☒
Factor X deficiency					☒
Factor XI deficiency					☒
Factor XIII deficiency					☒
Other hereditary bleeding disorders: type unknown					☒
Platelet disorders: Glanzmann's thrombasthenia					☒
Platelet disorders: Bernard Soulier Syndrome					☒
Platelet disorders: other or unknown					☒

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

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

Factor level below 5% ☒	Severe bleeding symptoms ☐	Other ☐ (please describe):	No data ☐

How are patients with von Willebrand Disease identified?

Laboratory diagnosis ☒	Severe bleeding symptoms ☐	Other ☐ (please describe):	No data ☐

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

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	421	146	300	31	☐
Hemophilia A female	0	0	0	0	☐
Hemophilia B male	34	42	50	7	☐
Hemophilia B female	0	0	0	0	☐

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
9	33	78	☐

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	Number with current inhibitors	No Data
Hemophilia A	13	☐
Hemophilia B	2	☐

11. Products used to treat hemophilia: What percentage of patients is treated with the following products?

Plasma	0 %
Cryoprecipitate	0 %
Plasma-derived concentrate	70 %
Recombinant concentrate	30 %
DDAVP (Desmopressin)	0 %

12. Products used to treat vWD: What percentage of patients is treated with the following products?

Plasma	0 %
Cryoprecipitate	0 %
Plasma-derived concentrate	100 %
DDAVP (Desmopressin)	0 %

13. HIV and hepatitis C infection among living people with hemophilia

Infectious Disease	Number of people infected	Percentage of people tested	No Data
HIV	2	77%	☐
Hepatitis C	183	97%	☐

14. HIV and hepatitis C infection among living people with von Willebrand disease

Infectious Disease	Number of people infected	Percentage of people tested	No Data
HIV	0	63%	☐
Hepatitis C	4	69%	☐

15. Number and cause of deaths of people with bleeding disorders (January 1-December 31, 2012)

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		

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?	7
How many **hemophilia comprehensive care centres** are there in your country?	4
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 children (under age 18) are on prophylaxis?	30	Precise: ☒ Estimate: ☐	Not known ☐
What percentage of adults (over age 18) are on prophylaxis?	21	Precise: ☒ Estimate: ☐	Not known ☐

D. The Cost and Use of Factor Concentrates

18. Annual usage of factor concentrates	**Factor VIII**	**Factor IX**	**Not known**
How many international units (IU) of factor concentrates were used in your country in 2012?	51 276 608	5 369 862	☐
How many international units of **plasma-derived** concentrates were used in your country in 2012?	32 973 006	4 615 700	☐
How many international units of **recombinant** concentrates were used in your country in 2012?	18 303 602	754 162	☐
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: 15%

19. Factor VIII Concentrates used in 2012
(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	10
☐	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	
☐	Emoclot D.I.	Kedrion	
☐	FACTANE	LFB	
☐	Factor 8 Y	BioProducts Lab.	
☐	Faktor VIII SDH Intersero	Intersero	
☒	Fanhdi	Grifols	8,5
☐	GreenEight	GreenCross	
☐	GreenGene	GreenCross	
☐	GreenMono	Greencross Corp	
☒	Haemate P (= Haemate HS)	CSL Behring	14
☒	Haemoctin SDH	Biotest	10

☐	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	
☒	Immunate	Baxter BioScience	10
☐	Koate DVI	Talecris	
☒	Kogenate FS = KOGENATE Bayer (in EU)	Bayer	10
☐	Monoclate P	CSL Behring	
☒	Octanate	Octapharma	10
☐	Octanativ-M	Octapharma	
☐	Optivate	Bio Products Laboratory	
☒	Recombinate rAHF	Baxter BioScience	10
☐	ReFacto AF	Pfizer (Wyeth)	
☐	Replenate	Bio Products Laboratory	
☒	Wilate	Octapharma	NA
☐	Xyntha	Pfizer (Wyeth)	
☐	Other:		

20. Factor IX Concentrates

(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	
☐	BeneFIX	Wyeth	
☐	Berinin-P = Berinin HS	CSL Behring	
☐	BETAFACT	LFB	
☐	Christmassin-M	Benesis	
☐	Factor IX Grifols	Grifols	
☐	Faktor IX SDN	Biotest	
☐	Hemo-B-RAAS	Shanghai RAAS	
☐	Haemonine	Biotest	
☒	Immunine	Baxter BioScience	11
☐	MonoFIX-VF	CSL Bioplasma	
☐	Mononine	CSL Behring	
☐	Nanotiv	Octapharma	
☐	Nonafact	Sanquin	

☐	Novact M	Kaketsuken	
☒	Octanine F	Octapharma	8,5
☐	Replenine – VF	BioProducts Lab.	
☐	Other:		

21. Prothrombin Complex Concentrates

(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	
☐	KASKADIL	LFB	
☒	Octaplex	Octapharma	10
☐	PPSB-human SD/Nano 300/600	German Red Cross NSTOB	
☐	Profilnine SD	Grifols	
☐	Proplex – T	Baxter BioScience	
☐	Prothrombinex- VF	CSL Bioplasma	
☒	Prothromplex-T	Baxter BioScience	10
☐	Prothroraas	Shanghai RAAS	
☐	UMAN Complex D.I.	Kedrion	
☐	Other:		

22. Other Products

(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
☐	Clottagen (fibrinogen)	LFB	
☐	Fibrinogen HT	Benesis	
☐	FIBRORAAS (fibrinogen)	Shanghai RAAS	
☒	Haemocomplettan P = Haemocomplettan HS (fibrinogen)	CSL Behring	
☐	Riastap	CSL Behring	
☒	Factor VII	Baxter BioScience	
☐	Factor VII	Bio Products	
☐	FACTEUR VII	LFB	
☐	Factor X P Behring	CSL Behring	
☐	Factor XI	Bio Products	
☒	HEMOLEVEN (Factor XI)	LFB	
☒	WILFACTIN (Von Willebrand Factor)	LFB	

☐	Fibrogammin P (=Fibrogammin HS) (Factor XIII)	CSL Behring	
☒	FEIBA	Baxter	
☒	NovoSeven (=Niastase) (activated factor VII)	NovoNordisk	Price: Vial size:
☐	Coagil 7 (activated factor VII)	Pharmstandard	Price: Vial size:
☐	Other:		

Completed by:
Date:
Contact info:
Please return to: Khalil Guliwala WFH Communications Assistant kguliwala@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.