A. National Hemophilia Organization

Organization name	Czech Society of Hemophilia
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Please Click Here **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** ☐ 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):

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)	1060	☐
2. Number of identified people with **von Willebrand disease** (VWD)	710	☐
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.)	65	☐
Do you consider these numbers to be accurate?	Yes ☒	Not sure ☐

Please click here 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	33	10	0	6
5 - 13 years old	103	15	0	43
14 - 18 years old	63	9	0	26
19 - 44 years old	414	51	0	341
45 years or older	311	51	0	294
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 5? (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	924	924	0	0	☐
Hemophilia B	136	136	0	0	☐
Hemophilia, type unknown	0	0	0	0	☐
von Willebrand disease	710	183	225	302	☐
Factor I deficiency	0				☐
Factor II deficiency	2	0	2		☐
Factor V deficiency	5	0	5		☐
Factor V+VIII deficiency	0				☐
Factor VII deficiency	26	11	15		☐
Factor X deficiency	3	1	2		☐
Factor XI deficiency	17	10	7		☐
Factor XIII deficiency	2	1	1		☐
Rare factor deficiency: type unknown	10	4	6		☐
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	435	147	305	37	☐
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
21	144	144	☐

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 2013	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	457	☒	☒	☐
Recombinant concentrate	203	☒	☒	☐
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	144	☒	☒	☐
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	794	☐
Hepatitis C	204	997	☐

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	188	☐
Hepatitis C	3	326	☐

15. Number and cause of deaths of people with bleeding disorders (January 1-December 31, 2013)

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?	7
How many **hemophilia comprehensive care centres** are there in your country?	2
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?	85	Precise: ☒ Estimate: ☐	Not known ☐
What percentage of eligible adults (over age 18) **with severe hemophilia** are on prophylaxis?	38	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 2013?	46 984 270	☐	5 539 509	☐
How many international units of **plasma-derived** concentrates were used in your country in 2013?	27 855 215	☐	4 547 576	☐
How many international units of **recombinant** concentrates were used in your country in 2013?	19 129 055	☐	991933	☐

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 2013

(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	
☐	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	
☐	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	
☒	Immunate	Baxter BioScience	
☐	Koate DVI	Talecris	
☒	Kogenate FS = KOGENATE Bayer (in EU)	Bayer	
☐	Monoclate P	CSL Behring	
☒	Octanate	Octapharma	
☐	Octanativ-M	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 2013

(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	
☐	MonoFIX-VF	CSL Bioplasma	
☐	Mononine	CSL Behring	
☐	Nanotiv	Octapharma	
☐	Nonafact	Sanquin	
☐	Novact M	Kaketsuken	
☒	Octanine F	Octapharma	
☐	Replenine – VF	BioProducts Lab.	
☐	Other:		

21. Prothrombin Complex Concentrates used in 2013

(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	
☐	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	
☐	Prothroraas	Shanghai RAAS	
☐	UMAN Complex D.I.	Kedrion	
☐	Other:		

22. Other Products used in 2013

(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:		

Please return to: globalsurvey@wfh.org
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or return by mail to: **World Federation of Hemophilia**
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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.