



# Dutch Hemophilia Registry

## Annual Report 2019



HemoNED Foundation  
May 2020

<https://hemoned.nl/en>

# Contents

<b>Introduction</b> .....	<b>3</b>
<b>Organisation</b> .....	<b>4</b>
<b>Results Dutch Hemophilia Registry</b> .....	<b>5</b>
General.....	<b>5</b>
Hemophilia.....	<b>8</b>
Diagnosis and demographic data.....	8
Viral infections.....	11
Inhibitors.....	11
Treatment.....	12
Von Willebrand disease.....	<b>14</b>
Diagnosis and demographic data.....	14
Inhibitors.....	15
Treatment.....	15
Other bleeding disorders.....	<b>16</b>
<b>Results VastePrik</b> .....	<b>17</b>
Diagnosis and demographic data.....	17
Infusions and bleeds.....	18
<b>Support</b> .....	<b>21</b>
<b>Contact</b> .....	<b>21</b>

# Introduction

This annual report describes data from the Dutch Hemophilia Registry and the VastePrik digital infusion log for home treatment available on 31 December 2019.

## Dutch Hemophilia Registry

The Dutch Hemophilia Registry (HemoNED Registry) was established in 2017 as a joint initiative from the Dutch Hemophilia Treaters Society (NVHB), the Dutch Hemophilia Patient Society (NVHP) and the Dutch Hemophilia Nurses Society (NVHV). For rare diseases like hemophilia, for which there is an effective but expensive treatment, a national hemophilia registry is an important tool for monitoring treatment and improving quality of care. The anonymized registry data are used for overview reports, and in the future for scientific research and drug safety studies.

## HemoNED foundation

The HemoNED Registry is managed by the HemoNED Foundation. The aim of the HemoNED Foundation is described as follows:

*“The Foundation aims to set up a nationwide registry of people with hemophilia and related disorders including data about their disease, treatment and complications, to perform scientific research, to publish reports and to provide education to contribute to an improvement of the quality of care.”*

The board of the Foundation has set up a **Steering Committee** that is responsible for assessing and approving the annual reports and data applications. In a Governance document the Steering Committee has described which parties are involved in the HemoNED project, their responsibilities and rights and how the management and access of the registry data is regulated.

## VastePrik

The digital infusion log VastePrik was launched in April 2018. VastePrik is available both as a mobile app and an online webpage, in order for participants to register their home treatment and bleeds that occur. VastePrik is mainly used by participants on prophylaxis. Both the participant and his/her treater have access to the logged infusions and bleeds through a secured online webpage; this way, they can evaluate and if necessary adapt the treatment course of the participant.

## Inclusion

All national certified Hemophilia Treatment Centers (HTCs) routinely invite possible participants for the Dutch Hemophilia Registry:

- Amsterdam UMC location AMC
- Erasmus MC Rotterdam
- LUMC Leiden & HagaZiekenhuis The Hague
- HTC NEM: Radboudumc Nijmegen & MUMC Maastricht & MMC Veldhoven/Eindhoven
- UMC Groningen
- UMC Utrecht (Van Creveld Clinic)

People with one of the following diagnoses will be eligible to participate in the HemoNED registry:

- Hemophilia A or B
- Carriers with hemophilia A or B, coagulation factor levels  $\leq 50\%$
- Von Willebrand disease, VWFact and/or VWFact and/or VWFrcf and /or FVIII levels  $\leq 30\%$ , and/or dependent on clotting factor concentrates
- Rare factor deficiencies and platelet disorders, prophylactic treatment and/or dependent on clotting factor concentrates at surgery/trauma
- Acquired hemophilia

At the start of the HemoNED Registry in 2017/2018 the priority was set to the inclusion of people with severe Hemophilia. In 2019 the inclusion was extended to people with other bleeding disorders. Health care providers from the HTC's manually complete the registry with relevant medical information. The registry database has built-in validation checks to ensure quality of data.

## Data analysis

The HemoNED project office analyzed the data on behalf of the Steering Committee. The statistical software SPSS was used to perform describing statistical analyses (crosstabs, bar charts etc.) to analyze and describe the data. The percentages shown are the valid percentages.

The HemoNED Foundation ensures that all information provided for research and publications is fully anonymized. To further prevent indirect traceability this annual report presents cells with values lower than 10 as '<10' or values have been aggregated with other (sub)categories.

## Publications

HemoNED presented a poster at the annual congress of the European Association of Hemophilia and Allied Disorders (Prague, 6-9 February 2019) entitled "Towards evaluation of hemophilia therapies in the Netherlands: a nationwide patient registry and digital infusion log" (<https://hemoned.nl/en/publications/publications-detail>). In collaboration with the Dutch Hemophilia Patient Society, HemoNED provided numbers for the Annual Global Survey 2018 of the World Federation of Hemophilia.

# Organisation

## In 2019 the HemoNED Foundation consisted of the following board members:

- **Chair: Dr. F.J.M. (Felix) van der Meer**, Internist LUMC
- **Secretary: Dr. M.H.E. (Mariëtte) Driessens**, Delegate Netherlands Hemophilia Patient Society (NVHP)
- **Treasurer: Dr. K. (Kathelijn) Fischer**, Pediatric hematologist Van Creveld Clinic UMC Utrecht

## The following representatives were part of the HemoNED Steering Committee in 2019:

- **Dr. F.J.M. (Felix) van der Meer**, chair Steering Committee Expertise center for hemophilia and related disorders LUMC - HagaZiekenhuis
- **Dr. K. (Kathelijn) Fischer**, Van Creveld Clinic UMC Utrecht
- **Prof. Dr. C.J. (Karin) Fijnvandraat**, Amsterdam UMC location AMC Hemophilia Treatment Center
- **Dr. M.J.H.A. (Marieke) Kruijff**, Erasmus MC Rotterdam Hemophilia Treatment Center
- **Dr. B.A.P. (Britta) Laros-van Gorkom**, Hemophilia Treatment Center Radboudumc Nijmegen, MUMC+ Maastricht & MMC Eindhoven/Veldhoven
- **Dr. R.Y.J. (Rienk) Tamminga**, UMC Groningen Hemophilia Treatment Center
- **Dr. J. (Jan) Schipper**, NVHP
- **N. (Nanda) Uitslager**, Dutch Hemophilia Nurses Society (NVHV)

## The members of the HemoNED Project Office were:

**Dr. G. (Geertje) Goedhart**, Project coordinator HemoNED, LUMC  
**Drs. E.M. (Liesbeth) Taal**, Data manager HemoNED, LUMC

# Results Dutch Hemophilia Registry

## General

Figure 1a Number of unique participants in the HemoNED registry by gender



### Total participants

Total completed **1907** (100%)



### Gender

Man **1529** (80%)

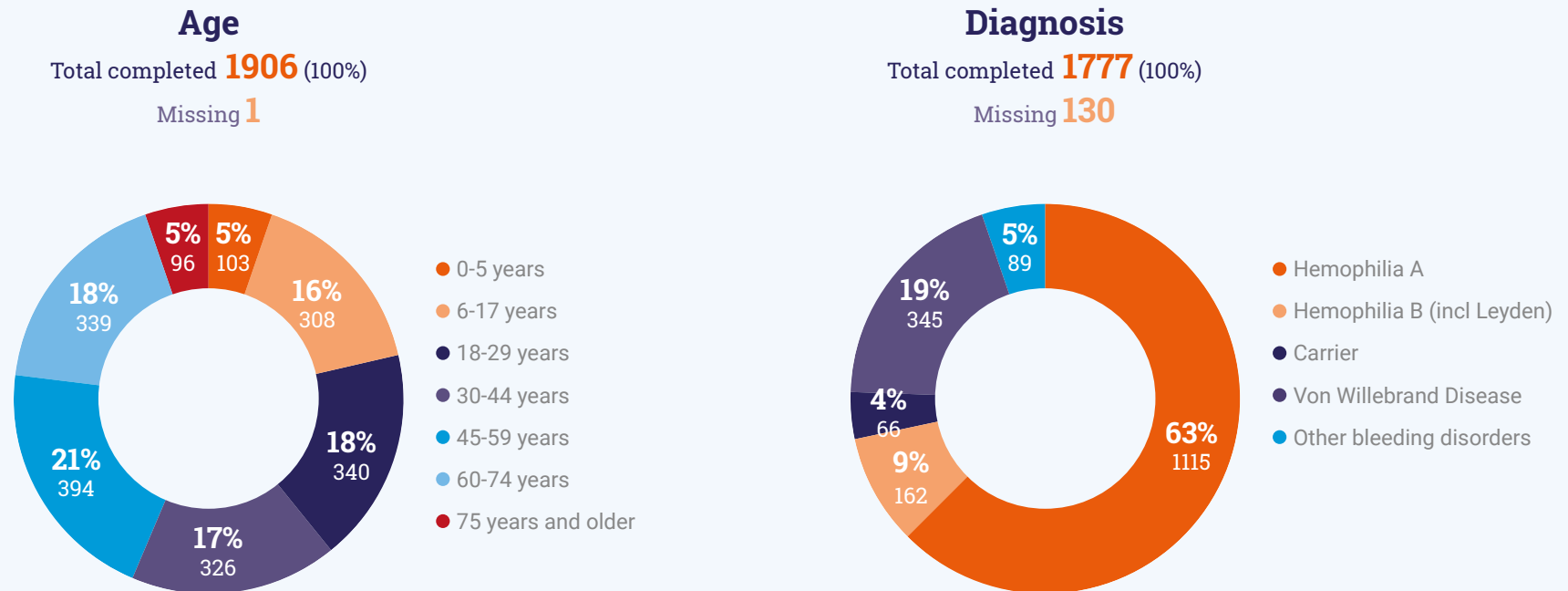
Woman **374** (20%)

Missing **4**

# Results Dutch Hemophilia Registry

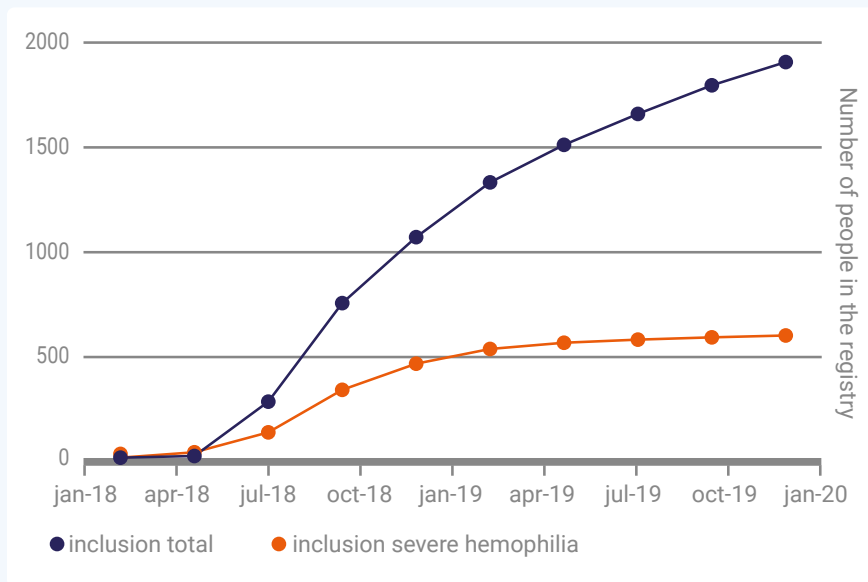
## General

Figure 1b Number of unique participants in the HemoNED registry by age and diagnosis

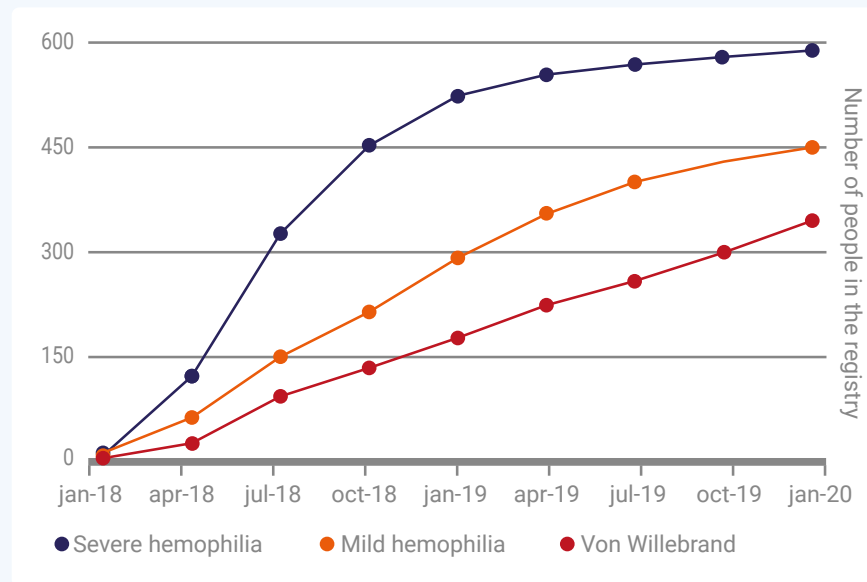


# General

**Figure 2a** Timeline: number of participants included in the HemoNED registry



**Figure 2b** Timeline: number of participants with severe hemophilia, mild hemophilia and von Willebrand disease included in HemoNED



# Hemophilia

## Diagnosis and demographic data

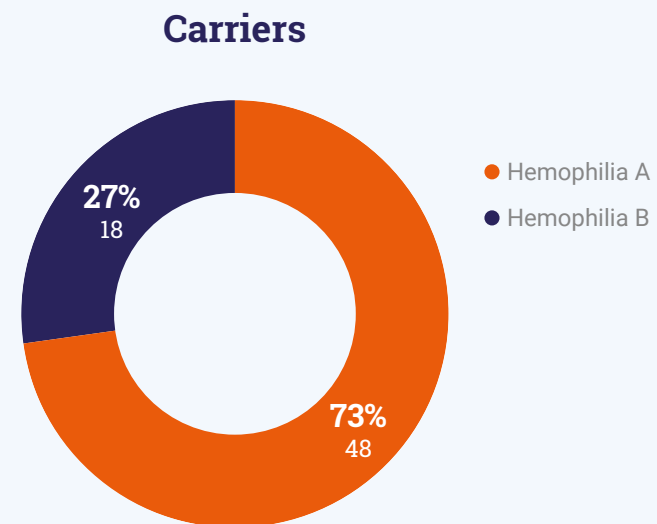
**Table 1** Number of participants in the HemoNED registry with diagnosis Hemophilia

Diagnosis	Number	%
<b>Hemophilia A</b>	<b>1115</b>	<b>100</b>
Severe	514	47
Moderate	168	15
Mild	411	38
Severity unknown	22	

Diagnosis	Number	%
<b>Hemophilia B</b>	<b>143</b>	<b>100</b>
Severe	75	54
Moderate	26	19
Mild	39	28
Severity unknown	3	

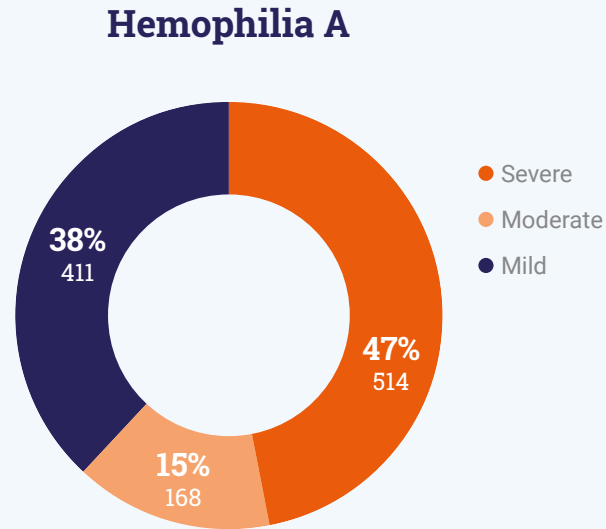
Diagnosis	Number	%
<b>Hemophilia B Leyden</b>	<b>19</b>	

**Figure 3** Carriers of Hemophilia A and B

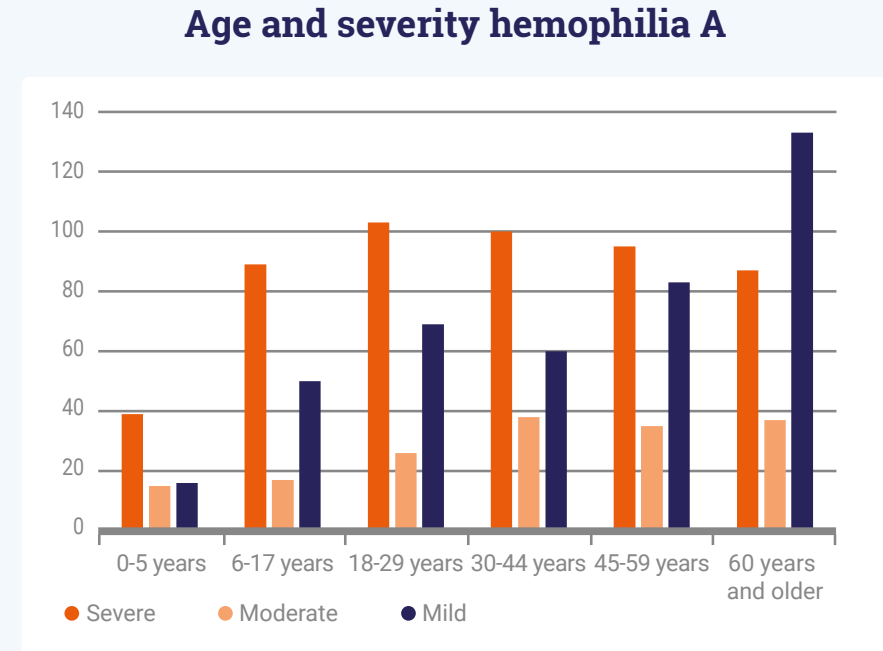


# Hemophilia

**Figure 4a** Participants with Hemophilia A by severity

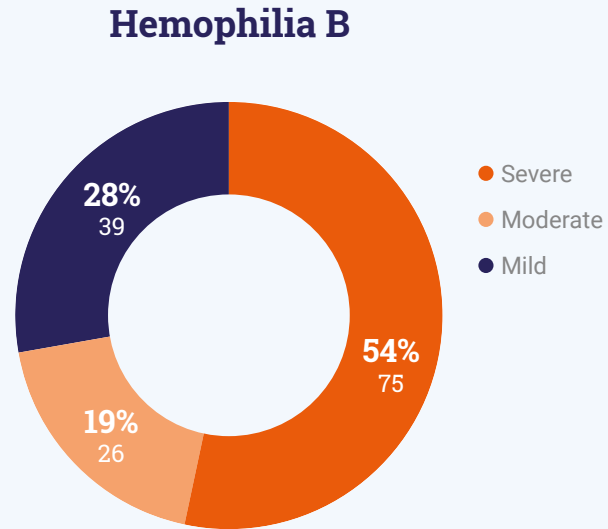


**Figure 4b** Participants with Hemophilia A by age and severity

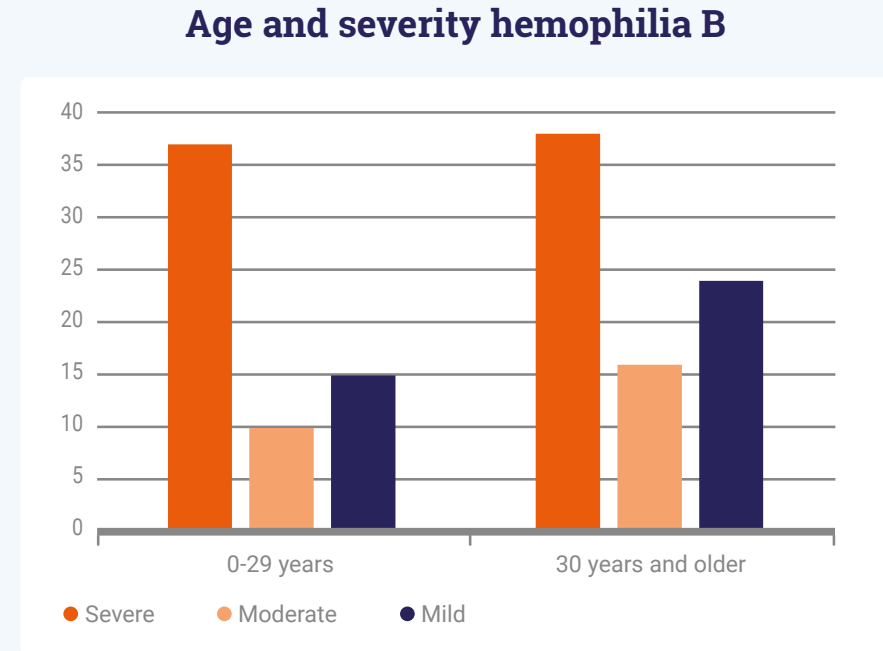


# Hemophilia

**Figure 5a** Participants with Hemophilia B by severity



**Figure 5b** Participants with Hemophilia B by age and severity



# Hemophilia

## Viral infections

**Table 2** Number of participants born before 1992 with diagnosis Hemophilia that suffer(ed) from a viral infection

Viral infection	Number	%
<b>Total completed</b>	<b>328</b>	<b>100</b>
Unknown	17	5
No	164	50
Yes*	147	45

Viral infection	Number	%
<b>HIV infection</b>	<b>&lt;10</b>	

Viral infection	Number	%
<b>Hepatitis B infection</b>	<b>45</b>	

Viral infection	Number	%
<b>Hepatitis C infection</b>	<b>134</b>	
Successfully treated	113	
Spontaneously cleared	<10	
Still infected	<10	
Unknown	<10	

\*Participants may (have) suffer(ed) from more than one infection

## Inhibitors

**Table 3** Inhibitor status of participants with diagnosis Hemophilia A or B

Inhibitors and Hemophilia A	Number	%
<b>Total completed</b>	<b>473</b>	<b>100</b>
Current inhibitor	16	3
In the past	43	9
Never	406	86
Unknown	<10	

Inhibitors and Hemophilia B	Number	%
<b>Total completed</b>	<b>72</b>	<b>100</b>
Current inhibitor	<10	
In the past	<10	
Never	71	99
Unknown	<10	

# Hemophilia

## Treatment

**Table 4** Number of participants with diagnosis moderate or severe Hemophilia A or B on prophylaxis

Prophylaxis	Number	%
<b>Hemophilia A Severe</b>		
<b>Total completed</b>	<b>252</b>	<b>100</b>
No	17	7
Yes	235	93
<b>Hemophilia A Moderate</b>		
<b>Total completed</b>	<b>95</b>	<b>100</b>
No	78	82
Yes	17	18
<b>Hemophilia B Severe</b>		
<b>Total completed</b>	<b>48</b>	<b>100</b>
No	0	0
Yes	48	100
<b>Hemophilia B Moderate</b>		
<b>Total completed</b>	<b>12</b>	<b>100</b>
No	<10	
Yes	<10	



# Hemophilia

**Table 5** Number of participants with diagnosis Hemophilia A or B by prescribed treatment product

Hemophilia A	Number	%
<b>Total completed</b>	<b>502</b>	<b>100</b>
Product A	211	42
Product B	105	21
Product C	54	11
Product D	50	10
Product E	23	4
Other products*	59	12

Hemophilia B	Number	%
<b>Total completed</b>	<b>80</b>	<b>100</b>
Product A	56	70
Product B	14	18
Other products*	10	12

\*Number of products too small (<10)

**Table 6** Number of participants with diagnosis Hemophilia A or B by type of product

Hemophilia A	Number	%
<b>Total completed</b>	<b>502</b>	<b>100</b>
Standard Half Life	434	86
Extended Half Life	27	5
Bypassing Agents	15	3
Plasma derived	<10	
Non Replacement Therapy	<10	
Other/combination of products	18	4

Hemophilia B	Number	%
<b>Total completed</b>	<b>80</b>	<b>100</b>
Standard Half Life	63	79
Extended Half life	16	20
Plasma derived	<10	

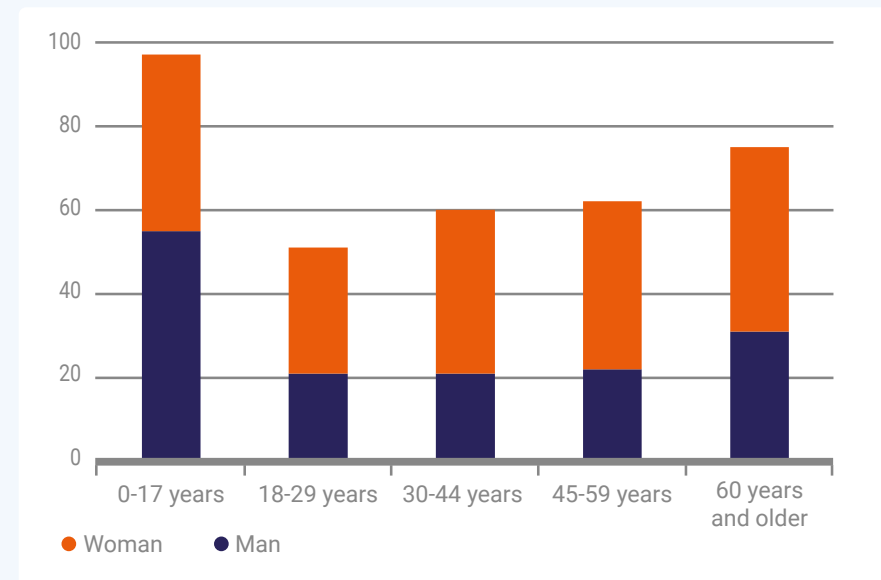
# Von Willebrand Disease

## Diagnosis and demographic data

**Table 7** Number of participants in HemoNED registry with diagnosis von Willebrand disease

	Number	%
<b>Von Willebrand</b>	<b>345</b>	<b>100</b>
Type 1	180	52
Type 2A	56	16
Type 2B	39	11
Type 2M	24	7
Type 3	27	8
Other/unknown	19	5

**Figure 6** Participants with von Willebrand disease by age and gender



# Von Willebrand Disease

## Inhibitors

**Table 8** Inhibitor status of participants with diagnosis von Willebrand disease

Inhibitors and von Willebrand disease	Number	%
<b>Total completed</b>	<b>135</b>	<b>100</b>
Current inhibitor	0	
In the past	0	
Never	125	93
Unknown	10	7

## Treatment

**Table 9** Number of participants with diagnosis von Willebrand disease by prescribed treatment product

Products and von Willebrand disease	Number	%
<b>Total completed</b>	<b>105</b>	<b>100</b>
Product A	94	90
Other products*	11	10

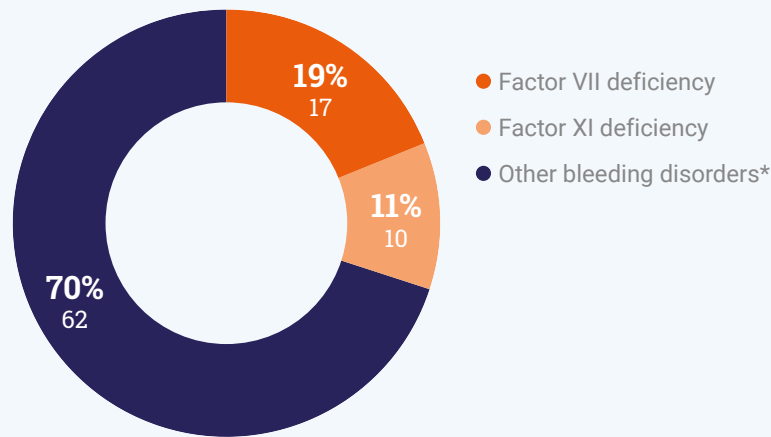
\*Number of products too small (<10)

# Other bleeding disorders

**Figure 7** Number of participants in HemoNED registry with other bleeding disorders

## Other bleeding disorder

Total completed **89** (100%)



\*Other bleeding disorders subcategories (<10 participants)

Other bleeding disorders in the HemoNED Registry:

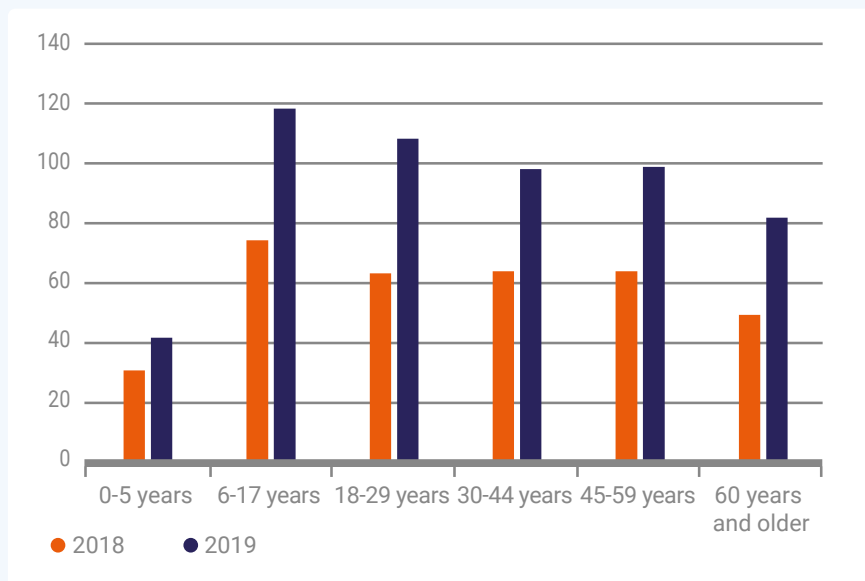
- Afibrinogenemia
- Dysfibrinogenemia
- Hypofibrinogenemia
- Hypodysfibrinogenemia
- Factor II deficiency
- Factor V deficiency
- Combined Factor V and Factor VIII deficiency
- Factor X deficiency
- Factor XIII deficiency
- Glanzmann's disease
- Storage Pool disease
- Alpha-2-antiplasmin deficiency
- Acquired deficiency
- Other factor deficiency
- Other platelets disorders



# Results VastePrik

## Diagnosis and demographic data

**Figure 8** Age distribution of the VastePrik users in 2018 (N=353) and 2019 (N=547)



**Table 10** Diagnosis of VastePrik users (usage ≥1)

Diagnosis	Number	%
<b>Total</b>	<b>547</b>	<b>100</b>
<b>Hemophilia A</b>	<b>414</b>	<b>79</b>
Severe	333	
Moderate	48	
Mild	22	
Severity unknown	11	
<b>Hemophilia B</b>	<b>65</b>	<b>12</b>
Severe	51	
Moderate	<10	
Mild	<10	
Severity unknown	<10	
<b>Von Willebrand</b>	<b>25</b>	<b>5</b>
Type 3	13	
Other types/unknown	12	
<b>Other bleeding disorders</b>	<b>19</b>	<b>3</b>
Missing	24	

## Infusions and bleeds

**Table 11** Number of infusions by reason reported in VastePrik (2018-2019)

Reason infusion	Number of infusions	%
Prophylaxis	47851	88
Precaution (risky activities)	1372	3
(Directly following a) Bleed	2657	5
Aftercare (after a bleed or surgery)	2388	4
<b>Total completed</b>	<b>54268</b>	<b>100</b>

**Table 12** Type of bleeds reported (2018-2019)

Type of bleed	Number of bleeds	%
Joint	1347	51
Muscle	528	20
Subcutaneous	185	7
Mucous membranes	138	5
Other	459	17
<b>Total</b>	<b>2657</b>	<b>100</b>

**Table 13** Bleed severity and cause of reported bleeds (2018-2019)

Bleed severity	Number of bleeds	%
Low	714	27
Average	1468	55
High	475	18
<b>Total</b>	<b>2657</b>	<b>100</b>

Cause	Number of bleeds	%
Spontaneously	1158	44
Overload	620	23
Accident or trauma	478	18
Postoperative	24	1
Other	377	14
<b>Total</b>	<b>2657</b>	<b>100</b>

**Table 14** Reported bleeds in VastePrik in 2019 (selection: regular VastePrik users on prophylaxis)

	Number of participants without bleeds	Number of participants with bleeds	Number of bleeds	A(J)BR**		Number of double bleeds*	A(J)BR without double bleeds	
				median (IQR)	range		median (IQR)	range
<b>All bleeds</b>	31	106	722	3 (1-7)	0 – 62	110	3 (1-6)	0 – 60
<b>Joint bleeds</b>	53	84	398	1 (0-4)	0 – 47	55	1 (0-3)	0 – 45

\* A double bleed is defined as a bleed logged at the same or next day and at the same location of the body as the previous bleed. These data are probably incorrect.

\*\*Annualized (Joint) Bleeding Rate

**Table 15** Most recently used product reported by VastePrik users with Hemophilia

	Number of users	%
<b>Hemophilia A</b>	<b>414</b>	<b>100</b>
Product A	152	37
Product B	22	5
Product C	70	17
Product D	63	15
Product E	53	13
Product F	26	6
Other products*	28	7


	Number of users	%
<b>Hemophilia B</b>	<b>65</b>	<b>100</b>
Product A	22	57
Product B	37	34
Other products*	<10	

\*Number of products too small (<10)

**Table 16** Most recently used product reported by VastePrik users with Hemophilia, by type of product

	Number of users	%
<b>Hemophilia A</b>	<b>414</b>	<b>100</b>
Standard Half Life	312	75
Extended Half Life	84	20
Bypassing Agents	<10	
Plasma derived	<10	
Non Replacement Therapy	<10	
Other	<10	

	Number of users	%
<b>Hemophilia B</b>	<b>65</b>	<b>100</b>
Standard Half Life	24	37
Extended Half Life	41	63

 [Back to contents](#)

## Support

HemoNED received a grant/research support from the following sponsors:

- Bayer B.V.
- CSL Behring B.V.
- Novo Nordisk B.V.
- Octapharma Benelux N.V.
- Pfizer B.V.
- Roche Nederland B.V.
- Shire Netherlands B.V. (part of Takeda)
- Swedish Orphan Biovitrum BVBA/SPRL

## Contact

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