NHD
U.K.H.C.D.O NATIONAL HAEMOPHILIA DATABASE

Bleeding Disorder Statistics for Scotland

April 2019 to March 2020

A report from the UK National Haemophilia Database

The following report is based on patients who are registered with the National Haemophilia Database with a Scottish postcode (unless otherwise stated), regardless of which Haemophilia Centre they were treated at.

Contents

New Registrations

Table 1 New registrations - Number of people newly registered between April 2019 & March 2020, by diagnosis and gender

Coagulation Defect	Male	Female	Total
Haemophilia A	14	0	14
Haemophilia A Carrier		12	12
Acquired Haemophilia A	5	6	11
Haemophilia B	5	0	5
Haemophilia B Carrier		5	5
von Willebrand disease	9	25	34
Acquired von Willebrand disease	2	0	2
F.V deficiency	2	2	4
F.VII deficiency	12	17	29
F.X deficiency	0	3	3
F.XI Deficiency	3	14	17
F.XIII Deficiency	0	1	1
Co-inherited diagnoses	2	3	5
Afibrinogenemia	1	0	1
Dysfibrinogenemia	6	13	19
Hypofibrinogenemia	2	5	7
Other Platelet Defects	4	4	8
Miscellaneous	0	1	1
Unclassified bleeding disorder	2	14	16
Total	**69**	**125**	**194**

Carrier of Haemophilia A includes and Females with FVIII deficiency
Carrier of Haemophilia B includes and Females with FIX deficiency and Factor IX Leyden carriers

Table 1 shows the number of new registrations to the National Haemophilia Database of people with a Scottish postcode.

Table 2 New registrations of haemophilia A & B between April 2019 & March 2020, by age and disease severity

Coagulation Defect	Age (years)	Number of Patients (factor level IU/dl)			
		< 1	1 - 5	> 5	Total
Haemophilia A	0 : 9	2	0	3	5
	10 : 19	0	2	0	2
	20 : 29	1	0	2	3
	30 : 39	0	1	1	2
	40 : 49	0	0	1	1
	50 : 59	0	0	1	1
	60 : 69	0	0	0	0
	70 +	0	0	0	0
	Total	**3**	**3**	**8**	**14**
Haemophilia B	0 : 9	0	0	1	1
	10 : 19	0	0	1	1
	20 : 29	0	1	1	2
	30 : 39	0	0	0	0
	40 : 49	0	0	0	0
	50 : 59	0	0	0	0
	60 : 69	0	0	1	1
	70 +	0	0	0	0
	Total	**0**	**1**	**4**	**5**

N.B Age calculated at mid-year, 30/09/2019

Table 2 shows the number of new registrations of people with Haemophilia A and B with a Scottish postcode. This is broken down by age and disease severity as per the ISTH severity classification.

In Register

Table 3 In register – The total number of people in register as of 31st March 2020 and the number treated between April 2019 & March 2020

Coagulation Defect	In Register				Treated (n)	Treated %
	Males	Females	Unknown	Total		
Haemophilia A	490	1	0	491	282	57.43%
Acquired Haemophilia A	18	27	1	46	9	20.00%
Haemophilia A Carrier		253	0	253	11	4.35%
Haemophilia A with Liver Transplant	4	0	0	4	0	0.00%
Haemophilia B	132	0	0	132	72	54.55%
Haemophilia B Carrier		73	0	73	6	8.22%
Haemophilia B with Liver Transplant	1	0	0	1	0	0.00%
von Willebrand disease	389	765	2	1,156	119	10.31%
Acquired von Willebrand Disease	10	11	2	23	2	9.52%
Probable von Willebrands disease	15	41	2	58	3	5.36%
Prothrombin Deficiency	1	2	0	3	0	0.00%
F.V deficiency	11	21	0	32	0	0.00%
F.VII deficiency	114	144	0	258	7	2.71%
F.X deficiency	15	33	0	48	1	2.08%
F.XI Deficiency	96	137	0	233	4	1.72%
F.XIII Deficiency	2	4	0	6	5	83.33%
Combined V+VIII Deficiency	1	2	0	3	2	66.67%
Co-inherited diagnoses	8	16	0	24	4	16.67%
Acquired F.V deficiency	0	1	0	1	0	0.00%
Acquired F.XIII Deficiency	0	1	0	1	0	0.00%
Acquired Deficiency (other)	1	0	0	1	0	0.00%
Afibrinogenemia	1	0	0	1	1	100.00%
Dysfibrinogenemia	89	145	0	234	2	0.85%
Hypofibrinogenemia	12	19	1	32	0	0.00%
Hypodysfibrinogenemia	6	5	0	11	1	9.09%
Glanzmann's Thrombasthenia	3	9	0	12	4	33.33%
Bernard Soulier	3	4	0	7	0	0.00%
Other platelet defects	61	153	1	215	2	0.93%
Miscellaneous	8	30	0	38	1	2.63%
Unclassified bleeding disorder	7	66	0	73	2	2.74%
Totals	**1,498**	**1,963**	**9**	**3,461**	**540**	

Table 3 shows the total number of active registrations of people with a Scottish postcode and the number who were issued treatment during 2019/20.

Table 4 In Register – The total number of people in the register as of 31st March 2020, by diagnosis and registered Haemophilia Centre

Coagulation Defect	Aberdeen	Dundee	Edinburgh	Glasgow	Inverness	Total
Haemophilia A	74	47	125	248	30	524
Haemophilia A Carrier	30	22	33	165	6	256
Haemophilia A with Liver Transplant	1	1	2	0	0	4
Acquired Haemophilia A	5	2	21	18	1	47
Haemophilia B	10	10	31	81	3	135
Haemophilia B Carrier	2	13	14	46	0	75
Haemophilia B with Liver Transplant	0	1	0	0	0	1
von Willebrand disease	164	189	166	633	41	1193
Acquired von Willebrands	1	2	11	10	0	24
Probable von Willebrands disease	1	33	7	17	0	58
F.V deficiency	6	0	3	23	0	32
F.VII deficiency	49	60	30	123	1	263
F.X deficiency	11	5	6	26	0	48
F.XI Deficiency	23	46	59	105	3	236
F.XIII Deficiency	3	0	1	2	0	6
Combined V+VIII Deficiency	0	0	1	2	0	3
Co-inherited diagnoses	1	7	2	14	0	24
Acquired F.V deficiency	0	0	0	1	0	1
Acquired F.XIII Deficiency	1	0	0	0	0	1
Acquired Deficiency (other)	0	0	0	1	0	1
Prothrombin Deficiency	0	0	2	1	0	3
Afibrinogenemia	0	0	0	1	0	1
Dysfibrinogenemia	5	20	36	174	1	236
Hypofibrinogenemia	6	0	6	19	2	33
Hypodysfibrinogenemia	0	0	10	1	0	11
Glanzmanns Thrombasthenia	1	3	5	3	0	12
Bernard Soulier	1	1	2	4	0	8
Other platelet defects	4	19	99	96	0	218
Miscellaneous	2	9	2	25	0	38
Unclassified bleeding disorder	0	2	3	69	0	74
Total	**401**	**492**	**677**	**1908**	**88**	**3,566**

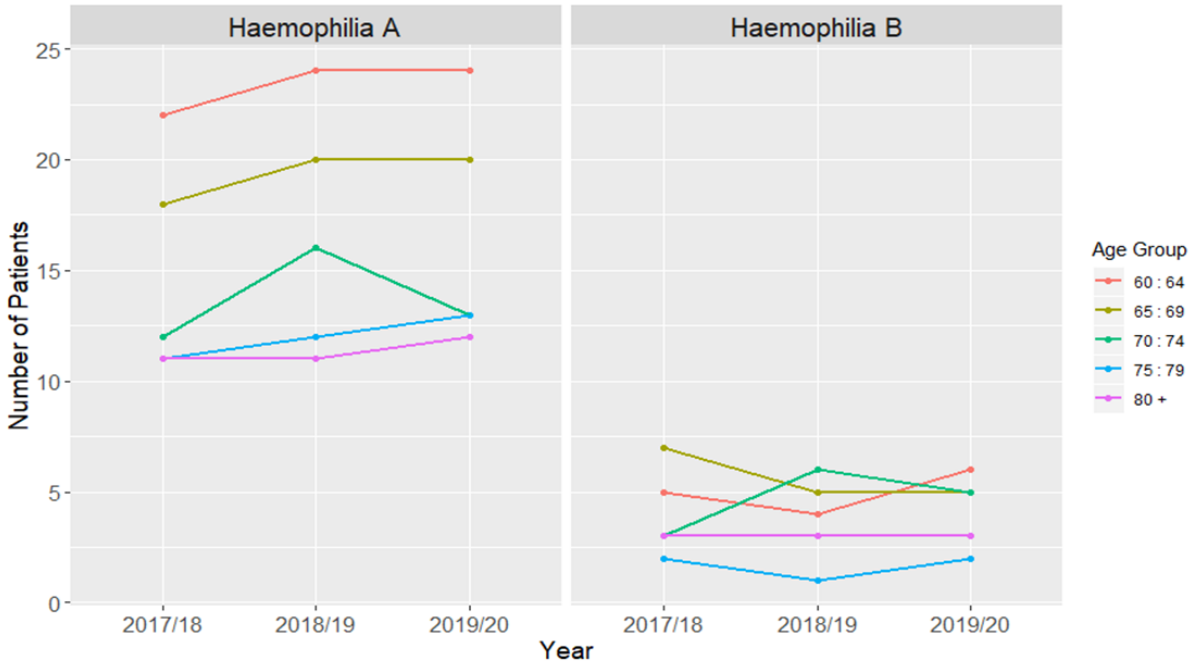
Table 4 shows the number of people registered at each Haemophilia Centre. Patients are allocated to their registered haemophilia centre regardless of their home postcode.

Table 5 In Register – The total number of people with Haemophilia A & B in the register as of 31st March 2020, by severity and age group

Coagulation Defect	Age (years)	Number of Patients (factor level iu/dl)			
		< 1	1 - 5	> 5	Total
Haemophilia A	<18 years	49	15	56	**120**
	≥18 years	95	54	222	**371**
	Total	**144**	**69**	**278**	**491**
Haemophilia B	<18 years	7	7	15	**29**
	≥18 years	20	35	48	**103**
	Total	**27**	**42**	**63**	**132**

Table 5 shows a more detailed breakdown of active registrations of people with a Scottish postcode with Haemophilia A and B. This is broken down by age and disease severity as per the ISTH severity classification.

Figure 1: Trend in people aged 60 years and above with Haemophilia A & B by age group



Data tables for figure 1

Haemophilia A					
Year	Age (years)	Number of Patients (factor VIII level iu/dl)			
		< 1	1 - 5	> 5	Total
2017/18	60 : 64	2	3	17	22
	65 : 69	3	3	12	18
	70 : 74	2	2	8	12
	75 : 79	1	1	9	11
	80 +	0	1	10	11
2018/19	60 : 64	3	3	18	24
	65 : 69	3	2	15	20
	70 : 74	2	2	12	16
	75 : 79	1	2	9	12
	80 +	0	1	10	11
2019/20	60 : 64	3	3	18	24
	65 : 69	3	2	15	20
	70 : 74	1	2	10	13
	75 : 79	1	1	11	13
	80 +	0	1	11	12

Haemophilia B					
Year	Age (years)	Number of Patients (factor IX level iu/dl)			
		< 1	1 - 5	> 5	Total
2017/18	60 : 64	1	0	4	5
	65 : 69	1	1	5	7
	70 : 74	0	2	1	3
	75 : 79	1	0	1	2
	80 +	0	1	2	3
2018/19	60 : 64	1	0	3	4
	65 : 69	1	1	3	5
	70 : 74	0	2	4	6
	75 : 79	1	0	0	1
	80 +	0	1	2	3
2019/20	60 : 64	1	0	5	6
	65 : 69	1	1	3	5
	70 : 74	0	2	3	5
	75 : 79	1	0	1	2
	80 +	0	1	2	3

Table 6 In Register – The number of people with selected rarer bleeding disorders in the register as of 31st March 2020 and the number treated between April 2019 & March 2020, by disease severity

Coagulation Defect	Number of Patients (factor level IU/dl)							
	<5		≥5		N/K		Total	
	In Reg	Treated	In Reg	Treated	In Reg	Treated	In Reg	Treated
F.V deficiency	1	0	31	0	0	0	32	0
F.VII deficiency	6	0	251	7	1	0	258	7
F.X deficiency	0	0	48	1	0	0	48	1
F.XI Deficiency	9	1	224	3	0	0	233	4
Total	**16**	**1**	**554**	**11**	**1**	**-**	**571**	**12**

Coagulation Defect	<2		2 - <10		≥ 10		Total	
	In Reg	Treated	In Reg	Treated	In Reg	Treated	In Reg	Treated
F.XIII Deficiency	2	2	2	2	2	1	6	5
Total	**2**	**2**	**2**	**2**	**2**	**1**	**6**	**5**

Table 6 shows the number of people with selected rarer bleeding disorders and a Scottish postcode known to the NHD during 2019/20. It is acknowledged that these rarer disorders have no recognised classification of disease severity. However, the table above gives an idea of the range of registered levels.

Table 7 In register – The total number of people with Von Willebrand Disease in the register as of 31st March 2020 and the number treated between April 2019 & March 2020, by disease severity, age group and gender

von Willebrand disease	VWD Activity IU/dl										Total	Treated
	<10	10 - 29	≥30	N/K	Sub Total	<10	10 - 29	≥30	N/K	Sub Total		
	<18 years					≥18 years						
Males												
Type 1	1	12	9	0	**22**	2	38	69	0	**109**	**131**	**10**
Type 2A	1	0	0	0	**1**	2	6	0	0	**8**	**9**	**3**
Type 2B	0	0	0	0	**0**	0	2	1	0	**3**	**3**	**1**
Type 2M	0	2	0	0	**2**	1	3	2	0	**6**	**8**	**2**
Type 2N	0	0	0	0	**0**	0	1	1	0	**2**	**2**	**1**
Type 2 Unspecified	1	0	0	0	**1**	2	1	5	0	**8**	**9**	**2**
Type 3	1				**1**	5				**5**	**6**	**4**
Type Unreported	5	21	26	0	**52**	12	46	100	0	**158**	**210**	**19**
Low VWF	0	0	3	0	**3**	0	0	8	0	**8**	**11**	**0**
										Sub Total Males	**389**	**42**
Females												
Type 1	0	10	5	0	**15**	4	78	182	0	**264**	**279**	**19**
Type 2A	1	0	0	0	**1**	7	8	1	0	**16**	**17**	**5**
Type 2B	0	1	1	0	**2**	0	4	4	0	**8**	**10**	**2**
Type 2M	1	2	0	0	**3**	8	13	0	0	**21**	**24**	**2**
Type 2N	0	0	0	0	**0**	1	2	1	0	**4**	**4**	**0**
Type 2 Unspecified	1	2	0	0	**3**	0	6	2	0	**8**	**11**	**1**
Type 3	1				**1**	3				**3**	**4**	**2**
Type Unreported	8	19	21	0	**48**	18	84	235	1	**338**	**386**	**36**
Low VWF	0	1	2	0	**3**	0	0	27	1	**28**	**31**	**8**
										Sub Total Females	**766**	**75**
										Grand Total - Males and Females	**1,155**	**117**

Table 7 shows people registered with von Willebrand disease broken down by age, activity level, subtype, and gender and treatment. Whilst there is no generally agreed severity classification for VWD, it is reported here in subdivisions from "<10, 10-29 and ≥30" to give some indication of the distribution of severity amongst the Scottish cohort.

Treatment

Table 8 People with a Scottish postcode, treated between April 2019 & March 2020 and region which issued the treatment, by diagnosis, all severities

Coagulation Defect	Region Issuing Treatment	Patients Treated (n)
Haemophilia A	East Anglia	1
	Scotland East	129
	Scotland West	154
	South Yorkshire & Bassetlaw	1
	Sub total	**285**
Haemophilia A Carrier	Scotland East	4
	Scotland West	7
	Sub total	**11**
Acquired Haemophilia A	Scotland East	2
	Scotland West	6
	Sub total	**8**
Haemophilia B	Scotland East	24
	Scotland West	49
	Sub total	**73**
Haemophilia B Carrier	Scotland East	1
	Scotland West	5
	Sub total	**6**
von Willebrand disease	Birmingham & Black Country	2
	Scotland East	57
	Scotland West	58
	Sub total	**117**
Probable von Willebrand disease	Scotland East	2
	Scotland West	1
	Sub total	**3**
Acquired von Willebrand Disease	Scotland East	2
	Sub total	**2**
F.VII deficiency	Scotland East	1
	Scotland West	6
	Sub total	**7**
F.X deficiency	Scotland West	1
		1
F.XI Deficiency	Scotland East	3
	Sub total	**3**
F.XIII Deficiency	Scotland East	4
	Scotland West	1
	Sub total	**5**

Continued overleaf....

Table 8 continued…

Coagulation Defect	Region	Patients Treated (n)
Combined V+VIII Deficiency	Scotland West	2
	Sub total	**2**
Co-inherited diagnoses	Scotland East	3
	Scotland West	1
	Sub total	**4**
Afibrinogenemia	Scotland West	1
		1
Dysfibrinogenemia	Scotland West	2
	Sub total	**2**
Hypodysfibrinogenemia	Scotland East	1
	Sub total	**1**
Glanzmann's Thrombasthenia	Scotland East	3
	Scotland West	1
	Sub total	**4**
Other Platelet defects	Scotland East	1
	Scotland West	1
	Sub total	**2**
Unclassified bleeding disorder	Scotland East	1
	Scotland West	1
		2
Miscellaneous	Scotland West	1
	Sub total	**1**
	Grand total	**540**

N.B This table may contain duplicates where a patient received treatment from more than one region

Table 8 reports people with a Scottish postcode by region which issued the treatment. Some people received treatment outside of Scotland. More detail on the treatment issued to people with severe Haemophilia A and B can be found in tables 12 and 18 respectively.

Table 9 People with a non-Scottish postcode, registered & treated at a Scottish Haemophilia Centre between April 2019 & March 2020, by diagnosis, all severities

Coagulation Defect	Patient's home postcode region	Patients Registered (n)	Patients Treated (n)
Haemophilia A	East of England	1	1
	London	4	3
	Midlands	1	1
	North East and Yorkshire	3	2
	North West	4	3
	South East	7	4
	South West	2	1
	Sub total	**22**	**15**
Haemophilia A Carrier	East of England	1	-
	Midlands	1	
	North East and Yorkshire	1	-
	Sub total	**3**	**-**
Haemophilia B	East of England	1	1
	Midlands	1	1
	North West	1	1
	Sub total	**3**	**3**
Haemophilia B Carrier	South East	1	-
	Sub total	**1**	**-**
von Willebrand disease	London	2	1
	Midlands	1	-
	North East and Yorkshire	9	1
	North West	6	2
	Northern Ireland	1	-
	South East	3	1
	Sub total	**22**	**5**
Acquired von Willebrands	South East	1	1
	Sub total	**1**	**1**
F.VII deficiency	North East and Yorkshire	3	-
	South East	1	-
	Sub total	**4**	**-**
F.XI Deficiency	South East	1	-
	South West	1	-
	Sub total	**2**	**-**
Dysfibrinogenemia	Midlands	1	-
Hypofibrinogenemia	London	1	-
Platelet Defects	Wales	0	-
	Sub total	**0**	**-**
	Grand total	**60**	**24**

The people reported in Table 9 were registered at or issued treatment from a Scottish Haemophilia Centre during 2019/20, however, they have a postcode recorded on the NHD which falls outside of Scotland.

Haemophilia A and Factor VIII use

Table 10 Factor VIII issued, by diagnosis

Coagulation Defect	Patients Treated (n)	FVIII (IU)			
		Plasma	Recombinant	Enhanced Half-Life	Total
Haemophilia A	258	485,500	44,796,600	5,080,000	50,362,100
Haemophilia A Carrier	8	-	41,000	-	41,000
Acquired Haemophilia A	5	-	107,000	-	107,000
Haemophilia B	1 - 2	-	1,000	-	1,000
von Willebrand disease	67	811,000	-	-	811,000
Acquired von Willebrands	1 - 2	95,000	-	-	95,000
Combined V+VIII Deficiency	1 - 2	-	12,000	-	12,000
Co-inherited diagnoses	1 - 2	2,500	-	-	2,500
Total	**338***	**1,394,000**	**44,957,600**	**5,080,000**	**51,431,600**

Includes products containing a combination of VWF and FVIII, which are reported in FVIII units

Table 10 shows the number of people with a Scottish postcode who were issued factor VIII concentrate during 2019/20. Also shown are the number of units issued, broken down by diagnosis and product type. No investigational FVIII was reported to have been issued.

Table 11 Products issued to treat Haemophilia A (including inhibitors)

Manufacturer	Product	Patients Treated (n)	Total Units
Bayer	Kogenate	1 - 2	68,000
Grifols	Fanhdi	1 - 2	485,500
Novo Nordisk	NovoEight	42	8,527,500
	NovoSeven (mg)	11	581
Octapharma	Nuwiq	1 - 2	60,000
Pfizer	ReFacto AF	96	16,903,350
Roche	Hemlibra (mg)	16	54,875
SOBI/Biogen	Elocta	20	5,080,000
Takeda	Advate	99	19,237,750
	FEIBA	3	1,161,000
	Desmopressin	16	444
	Investigational	1 - 2	720

Units in IU unless otherwise stated

Table 11 shows the number of units of products issued to people with a Scottish postcode with Haemophilia A, all severities, including those with inhibitors, broken down by supplier.

Table 12a Factor VIII issued to people with *Severe* Haemophilia A (incl. treatment for inhibitors), by issuing Haemophilia Centre

Haemophilia Centre Issuing Treatment	Severe Haemophilia A		
	Patients treated (n)	Total FVIII Units	Mean Usage
Aberdeen	21	6,151,000	292,905
Dundee	15	4,105,750	273,717
Edinburgh	27	7,969,250	295,157
Glasgow	68	16,627,100	244,516
Inverness	3	564,250	188,083

Table 12a reports the number of people with severe haemophilia A issued treatment during 2019/20 and the number of units of factor VIII issued. This is broken down by the haemophilia centre which issued the treatment.

Note: If a person is treated at multiple centres they are included for each centre.

Table 12b Factor VIII issued by region for people with *Severe* Haemophilia A (incl. treatment for inhibitors), by region

Region Issuing Treatment	Severe Haemophilia A		
	Patients treated (n)	Total FVIII Units	Mean Usage
Scotland East	66	18,790,250	284,701
Scotland West	68	16,627,100	244,516

Table 12b reports the number of people with severe haemophilia A treated and the number of units of factor VIII issued during 2019/20. This is broken down by region based on the person's registered haemophilia centre.

Note: If a person is treated at multiple centres they are included for each region.

Table 13 Factor VIII usage by Health Board for people with *Severe Haemophilia A* only (incl. treatment for inhibitors)

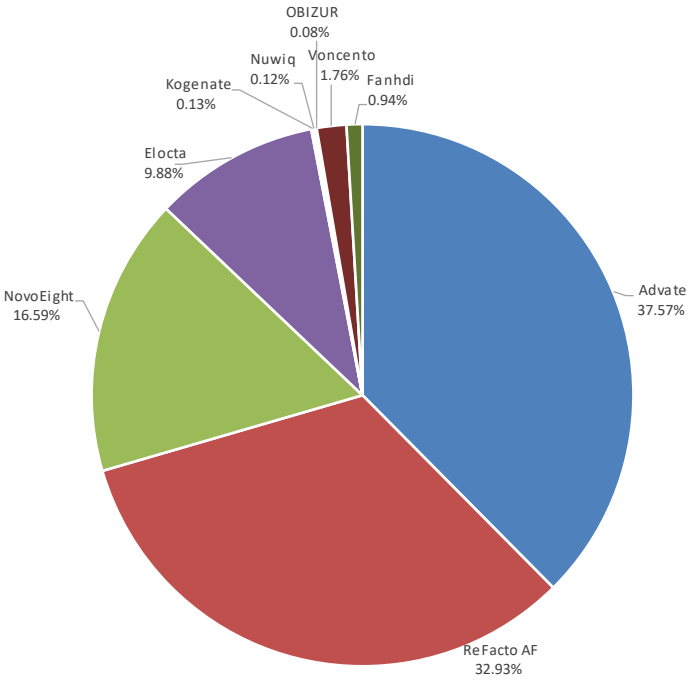
Health Board	General Population	Severe Haemophilia A			
		Patients treated (n)	Total FVIII Units (IU)	Mean Usage	FVIII Units Per Capita
Borders	115,510	2	1,795,000	897,500	15.54
Dumfries and Galloway	148,860	3	1,634,000	544,667	10.98
Western Isles	26,720	1	339,500	339,500	12.71
Grampian	585,700	19	6,091,250	320,592	10.40
Highland	321,700	6	1,842,500	307,083	5.73
Lanarkshire	661,900	6	1,670,000	278,333	2.52
Tayside	417,470	10	2,547,750	254,775	6.10
Ayrshire and Arran	369,360	11	2,795,250	254,114	7.57
Lothian	907,580	20	5,043,750	252,188	5.56
Fife	373,550	8	1,772,000	221,500	4.74
Greater Glasgow and Clyde	1,183,120	38	8,384,350	220,641	7.09
Shetland	22,920	3	642,250	214,083	28.02
Forth Valley	306,640	6	859,750	143,292	2.80
Scotland	**5,463,300**	**133**	**35,417,350**	**266,296**	**6.48**

Ranked by mean usage

Table 13 reports the number of people with severe haemophilia A issued treatment and number of units of factor VIII issued broken down by Health Board and ranked by the mean number of units issued per patient. Usage per capita of population is also reported.

Note: This table does not contain duplicates. People are allocated to a Health Board based on their home postcode.

Figure 2 Market share of factor VIII concentrates issued between April 2019 & March 2020



Includes products containing a combination of VWF and FVIII, which are reported in FVIII units
This pie chart is arranged in descending order of recombinant products by volume, then descending order of plasma products by volume

Manufacturer	Product	Units (IU)	Patients Treated (n)
Takeda	Advate	19,316,000	104
Pfizer	ReFacto AF	16,932,350	100
Novo Nordisk	NovoEight	8,530,250	44
SOBI/Biogen	Elocta	5,080,000	20
Bayer	Kogenate	68,000	1 - 2
Octapharma	Nuwiq	60,000	1 - 2
Shire	OBIZUR	39,000	4
CSL Behring	Voncento	907,500	68
Grifols	Fanhdi	485,500	1 - 2
	Total	**51,418,600**	**340***

** This is the total excluding numbers which have been suppressed.*
The table is arranged in descending order of recombinant products by volume, then descending order of plasma products by volume.

Figure 2 shows the market breakdown of factor VIII concentrates issued for all diagnoses, including people with inhibitors. Also included is a table showing the number of people issued with these products and the number of units issued.

NOTE: The number of people in this table cannot be compared with table 10 as this table includes people treated with more than one product type. The numbers of people treated in Table 10 do not contain duplicates.

Figure 3a Factor VIII units issued between April 2012 & March 2020 – all diagnoses, all severities, all people with a Scottish postcode

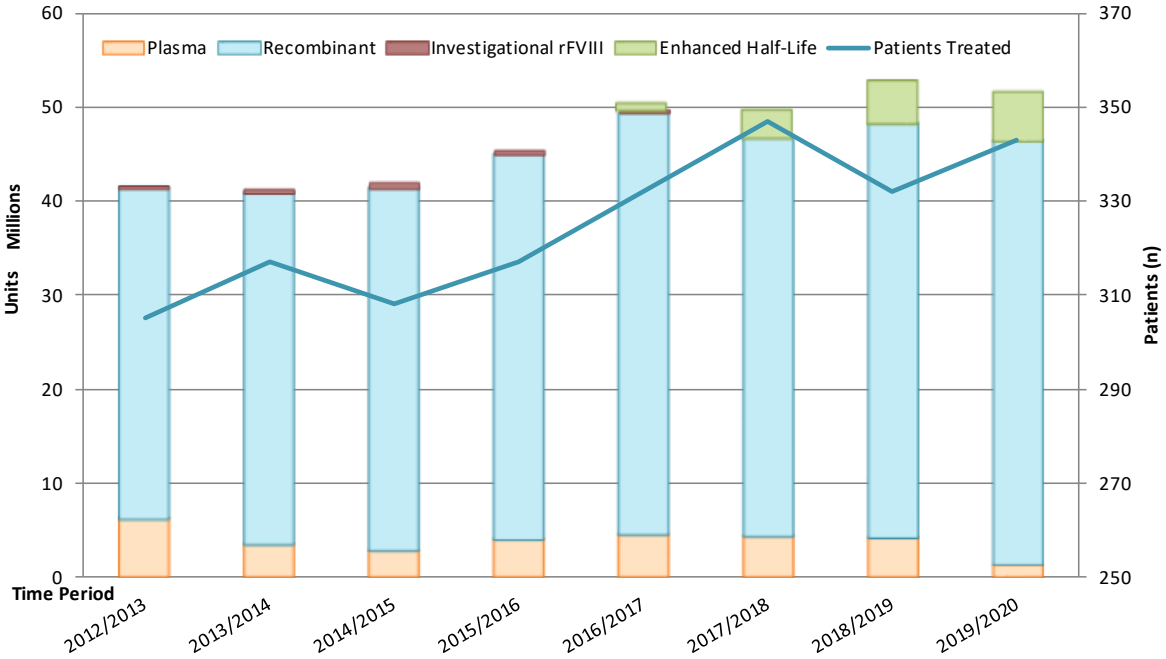


Figure 3b Factor VIII units issued between April 2012 & March 2020 – all diagnoses, all severities, people with a Scotland East postcode

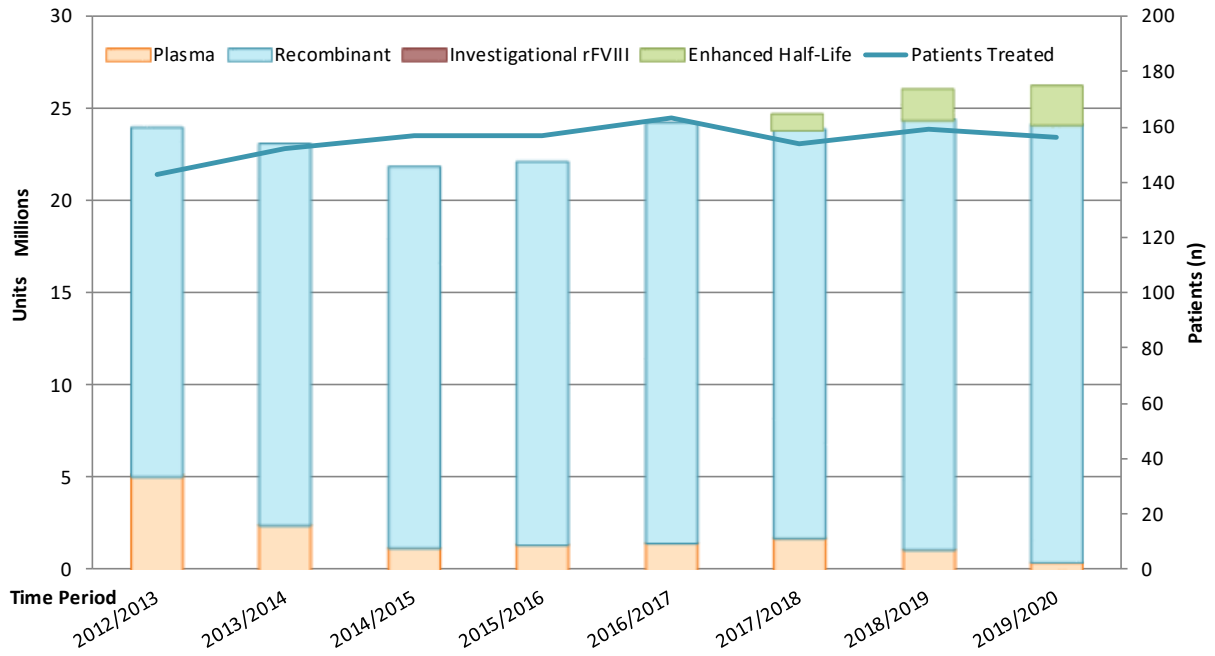
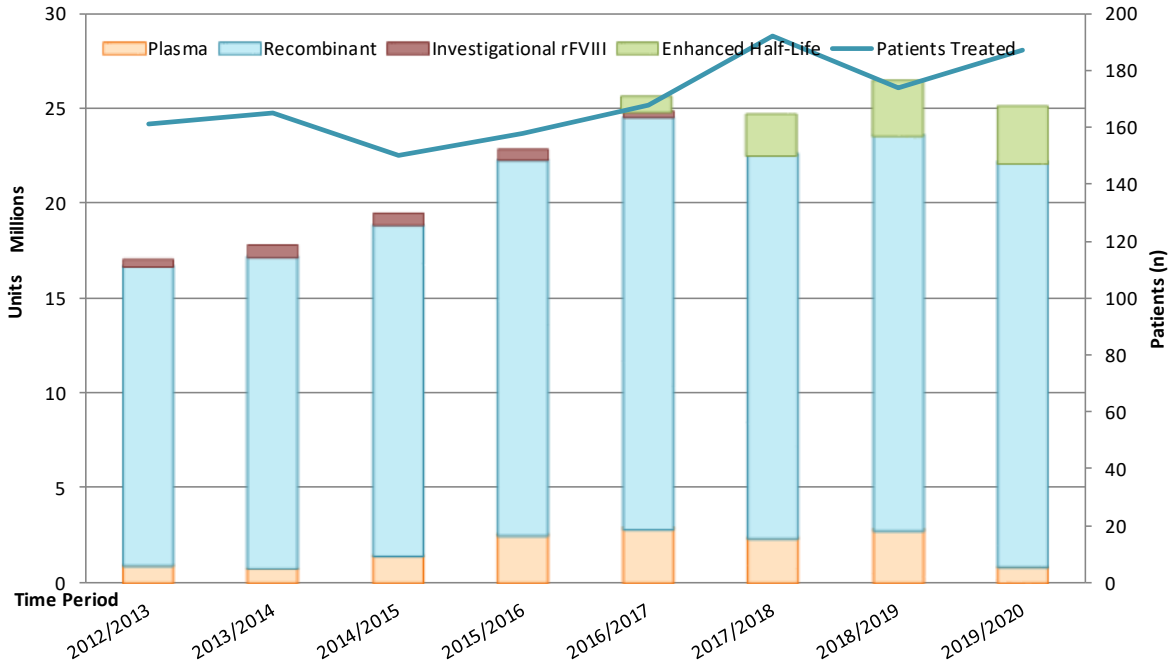


Figure 3c Factor VIII units issued between April 2012 & March 2020 – all diagnoses, all severities, people with a Scotland West postcode



Figures 3a to 3c include products containing a combination of VWF and FVIII, which are reported in FVIII units

Figures 3a to 3c give a historical view of the number of factor VIII units issued between 2012/13 and 2019/20 for all diagnoses and all severities. Figure 3a includes all people with a Scottish postcode. Figures 3b only includes people with a Scotland East postcode and Figure 3c those with a Scotland West postcode. The number of people treated is represented by the blue line using a secondary axis.

The data tables related to these graphs can be seen overleaf.

NOTE: Figure 3a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 3b and 3c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 3a is not directly comparable to Figures 3b and 3c as some people are dropped due to the fact that they were issued treatment from a haemophilia centre outside of Scotland. In addition, some people were issued treatment from both East and West Scotland.

Table 14a Data for Figure 3a - Factor VIII units issued between April 2012 & March 2020 – all diagnoses, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	6,192,350	1.00	35,082,895	1.00	472,202	1.00	0		41,747,447	1.00	305	1.00
2013/2014	3,399,100	0.55	37,403,895	1.07	630,000	1.33	0		41,432,995	0.99	317	1.04
2014/2015	2,819,500	0.46	38,464,250	1.10	674,750	1.43	0		41,958,500	1.01	308	1.01
2015/2016	3,986,050	0.64	40,798,250	1.16	613,500	1.30	0		45,397,800	1.09	317	1.04
2016/2017	4,402,000	0.71	44,793,250	1.28	330,500	0.70	849,750		50,375,500	1.21	332	1.09
2017/2018	4,231,000	0.68	42,418,250	1.21	0	0.00	3,089,750	1.00	49,739,000	1.19	347	1.14
2018/2019	4,076,500	0.66	44,054,750	1.26	0	0.00	4,580,950	1.48	52,712,200	1.26	332	1.09
2019/2020	1,394,000	0.23	44,957,600	1.28	0	0.00	5,080,000	1.64	51,431,600	1.23	343	1.12

Table 14b Data for Figure 3b - Factor VIII units issued between April 2012 & March 2020 – all diagnoses, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU		IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	5,060,350	1.00	18,898,895	1.00	0		0		23,959,245	1.00	143	1.00
2013/2014	2,499,100	0.49	20,586,645	1.09	0		0		23,085,745	0.96	152	1.06
2014/2015	1,273,000	0.25	20,551,500	1.09	0		0		21,824,500	0.91	157	1.10
2015/2016	1,390,500	0.27	20,753,250	1.10	0		0		22,143,750	0.92	157	1.10
2016/2017	1,478,500	0.29	22,761,000	1.20	0		0		24,239,500	1.01	163	1.14
2017/2018	1,795,500	0.35	22,060,250	1.17	0		919,000	1.00	24,774,750	1.03	154	1.08
2018/2019	1,186,000	0.23	23,198,750	1.23	0		1,720,000	1.87	26,104,750	1.09	159	1.11
2019/2020	465,500	0.09	23,638,750	1.25	0		2,106,500	2.29	26,210,750	1.09	156	1.09

Table 14c Data for Figure 3c - Factor VIII units issued between April 2011 & March 2020 – all diagnoses, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	884,000	1.00	15,731,500	1.00	472,202	1.00	0		17,087,702	1.00	161	1.00
2013/2014	744,000	0.84	16,388,750	1.04	630,000	1.33	0		17,762,750	1.04	165	1.02
2014/2015	1,385,500	1.57	17,428,250	1.11	674,750	1.43	0		19,488,500	1.14	150	0.93
2015/2016	2,472,050	2.80	19,693,000	1.25	613,500	1.30	0		22,778,550	1.33	158	0.98
2016/2017	2,785,500	3.15	21,612,250	1.37	330,500	0.70	849,750		25,578,000	1.50	168	1.04
2017/2018	2,319,500	2.62	20,204,500	1.28	0	0.00	2,170,750	1.00	24,694,750	1.45	192	1.19
2018/2019	2,746,500	3.11	20,790,500	1.32	0	0.00	2,860,950	1.32	26,397,950	1.54	174	1.08
2019/2020	777,500	0.88	21,315,100	1.35	0	0.00	2,973,500	1.37	25,066,100	1.47	187	1.16

Figure 4a Factor VIII units issued between April 2012 & March 2020 - *Severe Haemophilia A only*, all people with a Scottish postcode

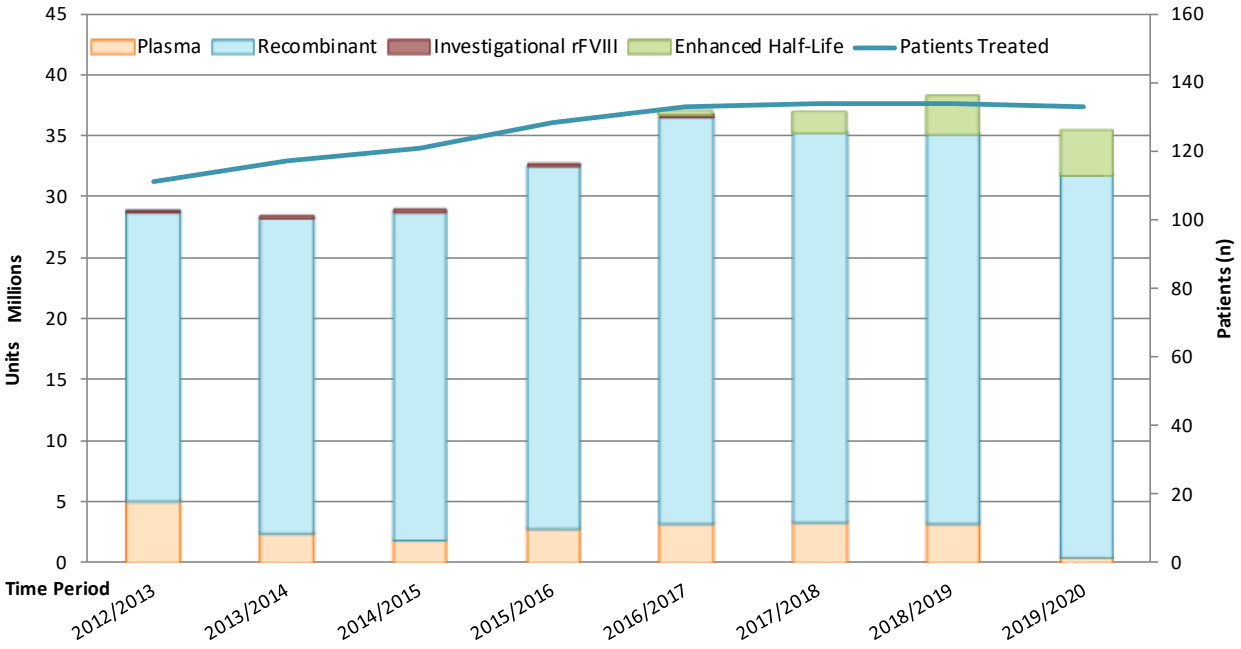


Figure 4b Factor VIII units issued between April 2012 & March 2020 - *Severe Haemophilia A only*, people with a Scotland East postcode

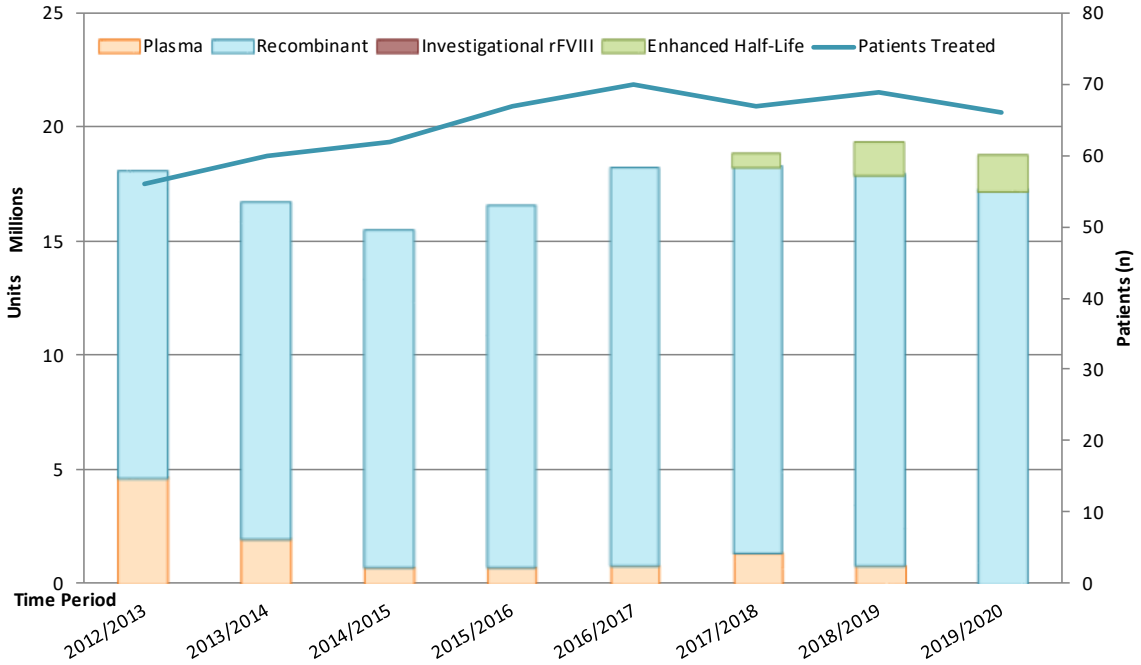
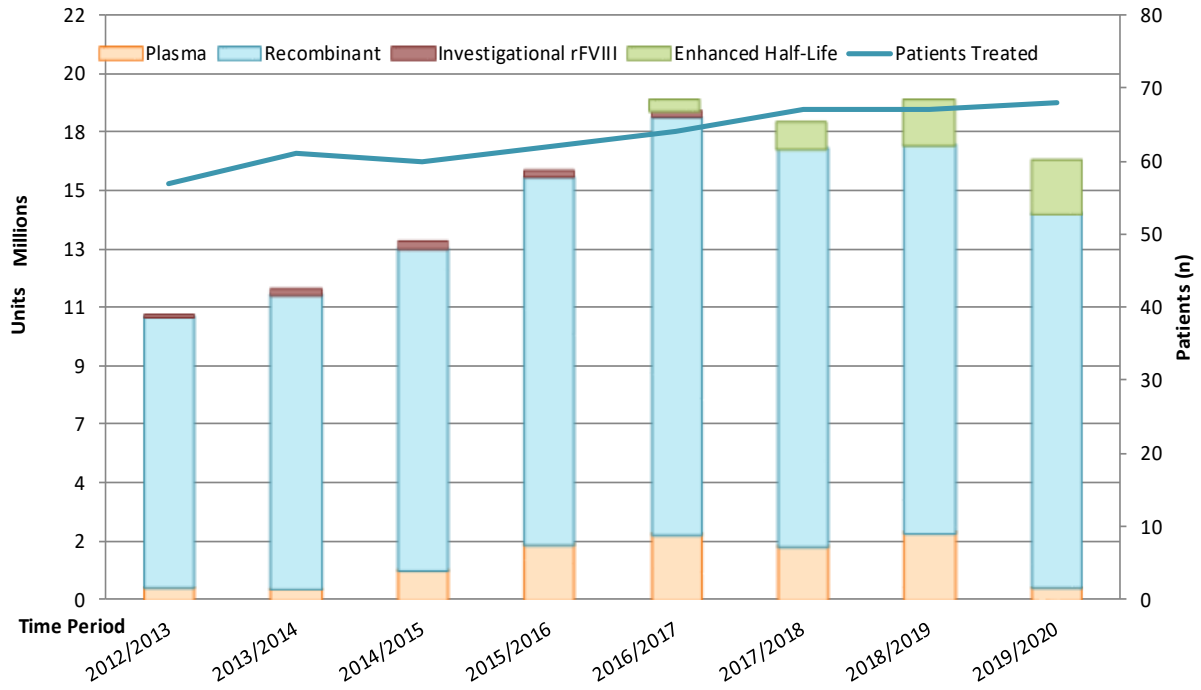


Figure 4c Factor VIII units issued between April 2012 & March 2020 – *Severe Haemophilia A only*, people with a Scotland West postcode



Figures 4a to 4c include products containing a combination of VWF and FVIII, which are reported in FVIII units

Figures 4a to 4c give a historical view of the number of factor VIII units issued between 2012/13 and 2019/20 for people with *severe haemophilia A only*. Figure 4a includes all patients with a Scottish postcode. Figures 4b includes only people with a Scotland East postcode and Figure 4c those with a Scotland West postcode. The number of people treated is represented by the blue line using a secondary axis.

The data tables related to these graphs can be seen overleaf.

NOTE: Figure 4a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 4b and 4c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 4a is not directly comparable to Figures 4b and 4c as some people are dropped due to the fact that they were issued treatment from a haemophilia centre outside of Scotland. In addition, some people were issued treatment from both East and West Scotland.

Table 15a Data for Figure 4a - Factor VIII units issued between April 2012 & March 2020 - *Severe Haemophilia A*, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	5,143,500	1.00	23,611,238	1.00	105,202	1.00	0		28,859,940	1.00	111	1.00
2013/2014	2,456,000	0.48	25,766,605	1.09	293,000	2.79	0		28,515,605	0.99	117	1.05
2014/2015	1,923,000	0.37	26,766,500	1.13	314,750	2.99	0		29,004,250	1.01	121	1.09
2015/2016	2,882,500	0.56	29,557,250	1.25	253,500	2.41	0		32,693,250	1.13	128	1.15
2016/2017	3,295,500	0.64	33,066,000	1.40	240,500	2.29	430,750		37,032,750	1.28	133	1.20
2017/2018	3,459,000	0.67	31,734,750	1.34	0	0.00	1,716,250	1.00	36,910,000	1.28	134	1.21
2018/2019	3,340,000	0.65	31,704,250	1.34	0	0.00	3,109,200	1.81	38,153,450	1.32	134	1.21
2019/2020	485,500	0.09	31,295,850	1.33	0	0.00	3,636,000	2.12	35,417,350	1.23	133	1.20

Table 15b Data for Figure 4b - Factor VIII units issued between April 2012 & March 2020 - *Severe Haemophilia A*, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU		IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	4,618,000	1.00	13,424,988	1.00	0				18,042,988	1.00	56	1.00
2013/2014	1,988,000	0.43	14,739,355	1.10	0				16,727,355	0.93	60	1.07
2014/2015	754,000	0.16	14,712,000	1.10	0				15,466,000	0.86	62	1.11
2015/2016	763,000	0.17	15,771,750	1.17	0				16,534,750	0.92	67	1.20
2016/2017	834,000	0.18	17,380,500	1.29	0				18,214,500	1.01	70	1.25
2017/2018	1,396,000	0.30	16,842,000	1.25	0		662,000	1.00	18,900,000	1.05	67	1.20
2018/2019	806,500	0.17	17,145,000	1.28	0		1,386,000	2.09	19,337,500	1.07	69	1.23
2019/2020	0	0.00	17,212,750	1.28	0		1,577,500	2.38	18,790,250	1.04	66	1.18

Table 15c Data for Figure 4c - Factor VIII units issued between April 2012 & March 2020 - *Severe Haemophilia A*, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	525,500	1.00	10,186,250	1.00	105,202	1.00	0		10,816,952	1.00	57	1.00
2013/2014	468,000	0.89	11,027,250	1.08	293,000	2.79	0		11,788,250	1.09	61	1.07
2014/2015	1,169,000	2.22	12,054,500	1.18	314,750	2.99	0		13,538,250	1.25	60	1.05
2015/2016	2,119,500	4.03	13,781,500	1.35	253,500	2.41	0		16,154,500	1.49	62	1.09
2016/2017	2,461,500	4.68	15,682,500	1.54	240,500	2.29	430,750		18,815,250	1.74	64	1.12
2017/2018	2,063,000	3.93	14,892,750	1.46	0	0.00	1,054,250	1.00	18,010,000	1.66	67	1.18
2018/2019	2,533,500	4.82	14,559,250	1.43	0	0.00	1,723,200	1.63	18,815,950	1.74	67	1.18
2019/2020	485,500	0.92	14,083,100	1.38	0	0.00	2,058,500	1.95	16,627,100	1.54	68	1.19

Haemophilia B and Factor IX use

Table 16 Factor IX issued, by diagnosis

Coagulation Defect	Patients Treated	FIX (IU)				
		Plasma	Recombinant	Enhanced Half-Life	Investigational	Total
Haemophilia B	71	340,000	4,123,750	2,540,250	-	7,004,000
Haemophilia B Carrier	6	-	23,500	-	-	23,500
Total	**77**	**340,000**	**4,147,250**	**2,540,250**	**-**	**7,027,500**

Table 16 shows the number of people with a Scottish postcode who were issued factor IX concentrate during 2019/20. Also shown are the number of units issued broken down by diagnosis and product type.

Table 17 Products issued to treat Haemophilia B (including inhibitors)

Manufacturer	Product	Patients Treated (n)	Total Units
CSL Behring	IDELVION	21	1,983,000
Grifols	Alphanine	1 - 2	340,000
Novo Nordisk	NovoSeven (mg)	1 - 2	450
	Refixia	1 - 2	207,000
Pfizer	BeneFIX	48	4,123,750
	ReFacto AF	1 - 2	1,000
SOBI/Biogen	ALPROLIX	4	350,250
	Investigational	1 - 2	560

Units in IU unless otherwise stated

Table 17 shows the number of units of products issued to people with a Scottish postcode with Haemophilia B, all severities, including those with inhibitors, broken down by supplier.

Table 18a Factor IX issued to people with *Severe* Haemophilia B (incl. treatment for inhibitors), by issuing Haemophilia Centre

Haemophilia Centre Issuing Treatment	Severe Haemophilia B		
	Patients treated (n)	Total FIX Units	Mean Usage
Aberdeen	4	290,500	72,625
Dundee	5	456,000	91,200
Edinburgh	1	6,000	6,000
Glasgow	17	2,520,500	148,265

Table 18 reports the number of people with severe haemophilia B treated and the number of units of factor IX issued during 2019/20. This is broken down by the haemophilia centre which issued the treatment.

Note: If a person is treated at multiple centres they are included for each centre.

Table 18b Factor IX issued to people with *Severe* Haemophilia B (incl. treatment for inhibitors), by region

Region Issuing Treatment	Severe Haemophilia B		
	Patients treated (n)	Total FIX Units	Mean Usage
Scotland East	10	752,500	75,250
Scotland West	17	2,520,500	148,265

Table 18b reports the number of people with severe haemophilia B treated and the number of units of factor IX issued during 2019/20. This is broken down by region based on the person's postcode as recorded on the NHD.

Note: If a person is treated at multiple centres they are included for each centre.

Table 19 Factor IX usage by Health Board for people with *Severe Haemophilia B* only (incl. treatment for inhibitors)

Health Board	General Population	Severe Haemophilia B			
		Patients treated (n)	Total FIX Units (IU)	Mean Usage	FIX Units Per Capita
Forth Valley	306,640	1	513,000	513,000	1.67
Ayrshire and Arran	369,360	2	592,000	296,000	1.60
Greater Glasgow and Clyde	1,183,120	8	911,750	113,969	0.77
Lanarkshire	661,900	4	407,500	101,875	0.62
Tayside	417,470	5	456,000	91,200	1.09
Grampian	585,700	4	290,500	72,625	0.50
Highland	321,700	2	102,250	51,125	0.32
Borders	115,510	0	-	-	-
Dumfries and Galloway	148,860	0	-	-	-
Fife	373,550	0	-	-	-
Lothian	907,580	0	-	-	-
Shetland	22,920	0	-	-	-
Western Isles	26,720	0	-	-	-
Scotland	**5,463,300**	**26**	**3,273,000**	**125,885**	**0.60**

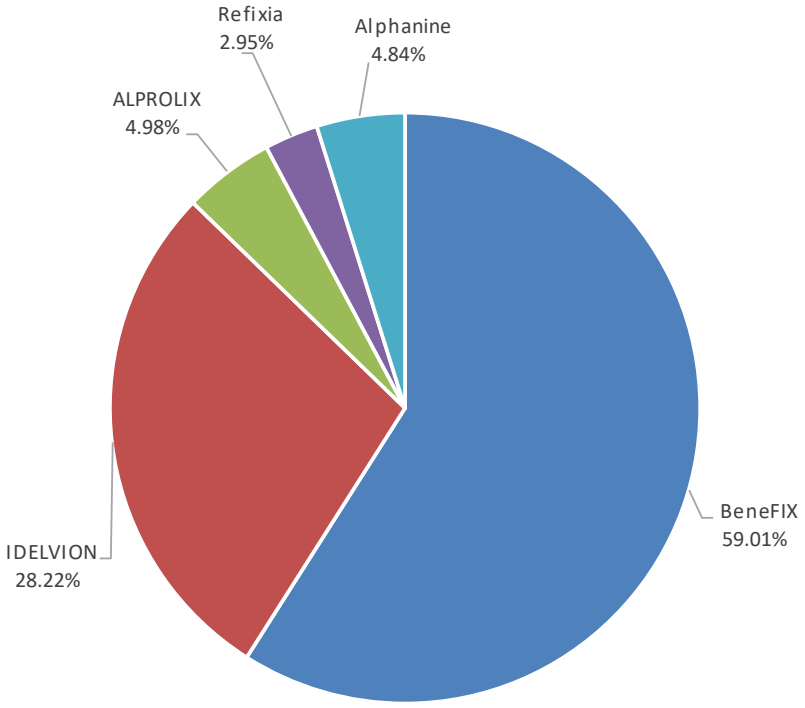
Ranked by mean usage

Mid-2019 population estimates Scotland supplied by National Records of Scotland under the Open Government Licence v3.0 © Crown Copyright 2020

Table 19 reports the number of people with severe haemophilia B treated and number of units of factor IX issued broken down by Health Board and ranked by the mean number of units issued per person. Usage per capita of population is also reported.

This table does not contain duplicate numbers of people. People are allocated to a Health Board based on their home postcode.

Figure 5 Market share of factor IX concentrates issued to people with a Scottish postcode between April 2019 & March 2020



This pie chart is arranged in descending order of recombinant products by volume, then descending order of plasma products by volume

Manufacturer	Product	Units (IU)	Patients Treated (n)
Pfizer	BeneFIX	4,147,250	53
CSL Behring	IDELVION	1,983,000	21
SOBI/Biogen	ALPROLIX	350,250	4
NovoNordisk	Refixia	207,000	1 - 2
Grifols	Alphanine	340,000	1 - 2
	Total	**7,027,500**	**78***

** This is the total excluding numbers which have been suppressed.*

The table is arranged in descending order of recombinant products by volume, then descending order of plasma products by volume.

Figure 5 shows the market breakdown of factor IX concentrates issued for all diagnoses, including people with inhibitors. Also included is a table showing the number of people issued with these products and the number of units issued.

NOTE: The number of people in this table cannot be compared with table 16 as this table includes people treated with more than one product type. The patient numbers in Table 16 do not contain duplicates.

Figure 6a Factor IX units issued between April 2012 & March 2020 – all diagnoses, all severities, all people with a Scottish postcode

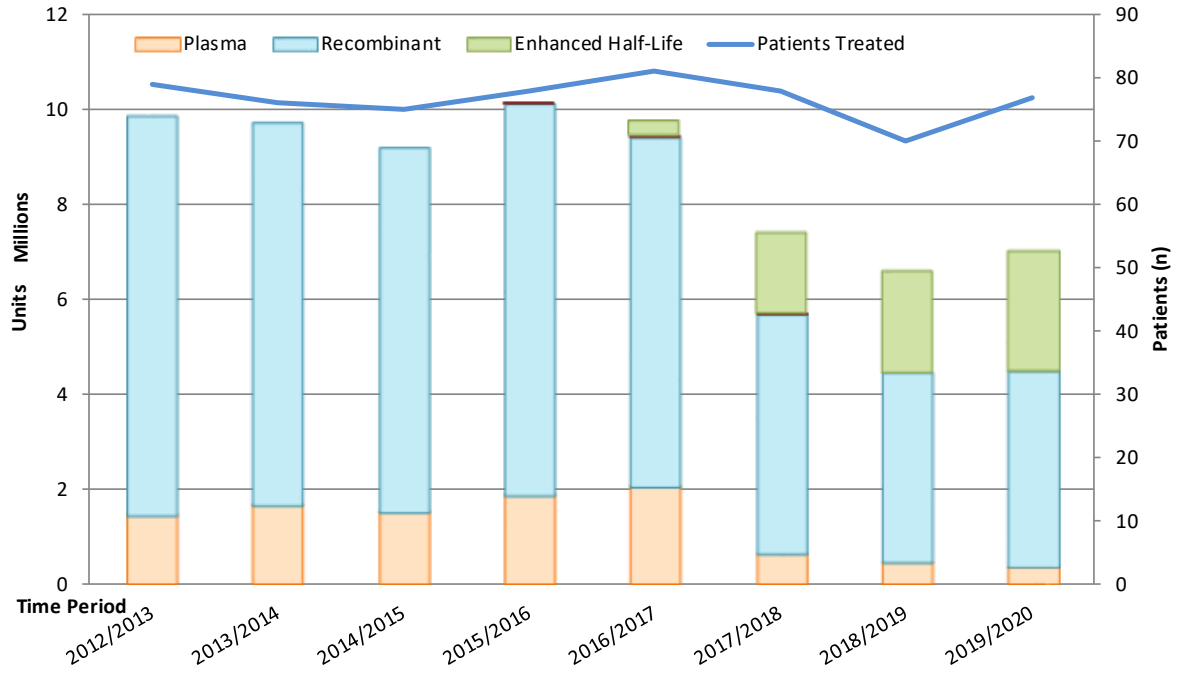


Figure 6b Factor IX units issued between April 2012 & March 2020 – all diagnoses, all severities, people with a Scotland East postcode

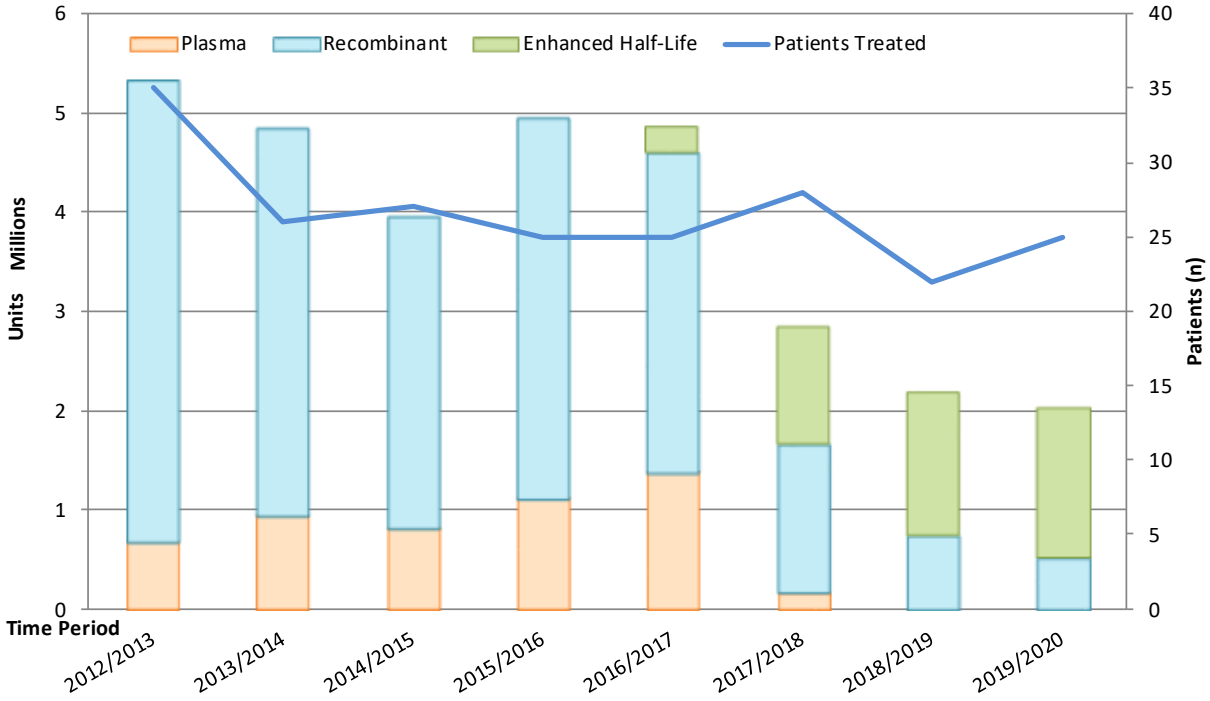
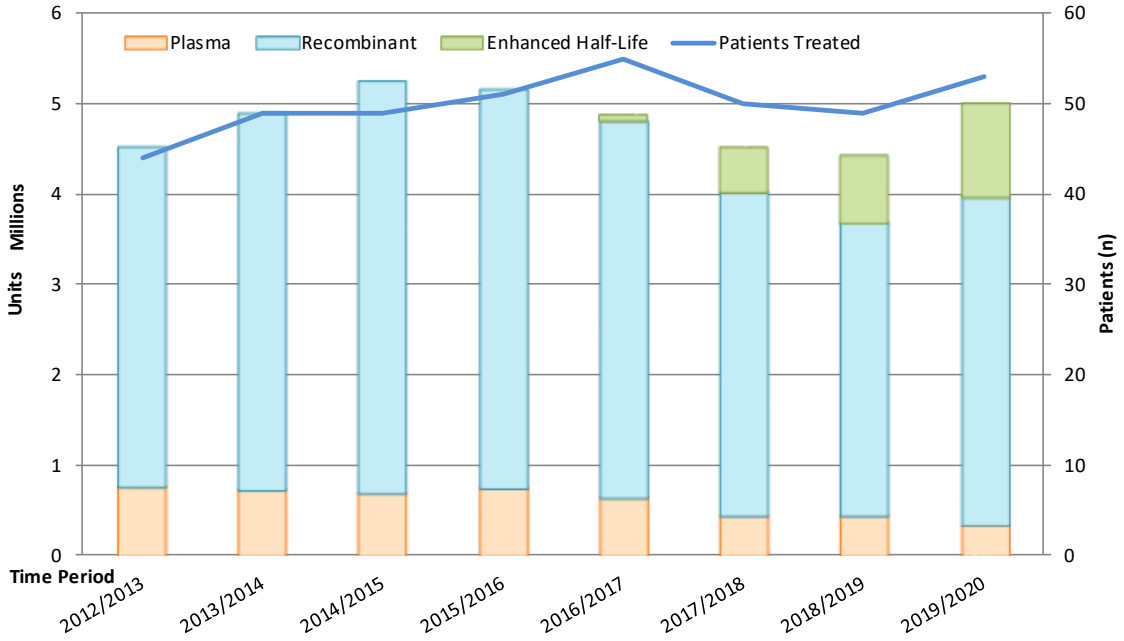


Figure 6c Factor IX units issued between April 2012 & March 2020 – all diagnoses, all severities, people with a Scotland West postcode



Figures 6a to 6c give a historical view of the number of factor IX units issued between 2012/13 and 2019/20 for all diagnoses and all severities. Figure 6a includes all people with a Scottish postcode. Figures 6b includes only people with a Scotland East postcode and Figure 6c those with a Scotland West postcode. The number of people treated is represented by the blue line using a secondary axis.

The data tables related to these graphs can be seen overleaf.

NOTE: Figure 6a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 6b and 6c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 6a is not directly comparable to Figures 6b and 6c as some people are dropped due to the fact that they were issued treatment from a haemophilia centre outside of Scotland. In addition, some patients were issued treatment from both East and West Scotland.

Table 20a Data for figure 6a - Factor IX units issued between April 2012 & March 2020 – all diagnoses, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2016/17	IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	1,426,250	1.00	8,449,486	1.00	0		0		9,875,736	1.00	79	1.00
2013/2014	1,656,935	1.16	8,071,089	0.96	0		0		9,728,024	0.99	76	0.96
2014/2015	1,501,780	1.05	7,685,500	0.91	0		0		9,187,280	0.93	75	0.95
2015/2016	1,862,370	1.31	8,248,000	0.98	2,500		0		10,112,870	1.02	78	0.99
2016/2017	2,013,200	1.41	7,381,000	0.87	47,653	1.00	325,500		9,767,353	0.99	81	1.03
2017/2018	614,305	0.43	5,068,750	0.60	25,588	0.54	1,692,500	1.00	7,401,143	0.75	78	0.99
2018/2019	441,000	0.31	3,999,250	0.47	0	0.00	2,172,250	1.28	6,612,500	0.67	70	0.89
2019/2020	340,000	0.24	4,147,250	0.49	0	0.00	2,540,250	1.50	7,027,500	0.71	77	0.97

Table 20b Data for figure 6b - Factor IX units issued between April 2012 & March 2020 – all diagnoses, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU		IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	667,250	1.00	4,666,236	1.00	0		0		5,333,486	1.00	35	1.00
2013/2014	928,935	1.39	3,906,589	0.84	0		0		4,835,524	0.91	26	0.74
2014/2015	802,780	1.20	3,143,500	0.67	0		0		3,946,280	0.74	27	0.77
2015/2016	1,113,870	1.67	3,831,000	0.82	0		0		4,944,870	0.93	25	0.71
2016/2017	1,366,700	2.05	3,221,500	0.69	0		261,000		4,849,200	0.91	25	0.71
2017/2018	171,805	0.26	1,487,000	0.32	0		1,181,500	1.00	2,840,305	0.53	28	0.80
2018/2019	0	0.00	748,750	0.16	0		1,436,000	1.22	2,184,750	0.41	22	0.63
2019/2020	0	0.00	507,500	0.11	0		1,527,500	1.29	2,035,000	0.38	25	0.71

Table 20c Data for Figure 6c - Factor IX units issued between April 2012 & March 2020 - all diagnoses, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU		IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	759,000	1.00	3,769,750	1.00	0		0		4,528,750	1.00	44	1.00
2013/2014	728,000	0.96	4,159,500	1.10	0		0		4,887,500	1.08	49	1.11
2014/2015	699,000	0.92	4,542,000	1.20	0		0		5,241,000	1.16	49	1.11
2015/2016	748,500	0.99	4,411,000	1.17	0		0		5,159,500	1.14	51	1.16
2016/2017	646,500	0.85	4,159,500	1.10	0		64,500		4,870,500	1.08	55	1.25
2017/2018	442,500	0.58	3,581,750	0.95	0		502,750	1.00	4,527,000	1.00	50	1.14
2018/2019	441,000	0.58	3,250,500	0.86	0		736,250	1.46	4,427,750	0.98	49	1.11
2019/2020	340,000	0.45	3,639,750	0.97	0		1,012,750	2.01	4,992,500	1.10	53	1.20

Figure 7a Factor IX units issued between April 2012 & March 2020 - *Severe Haemophilia B only*, all people with a Scottish postcode

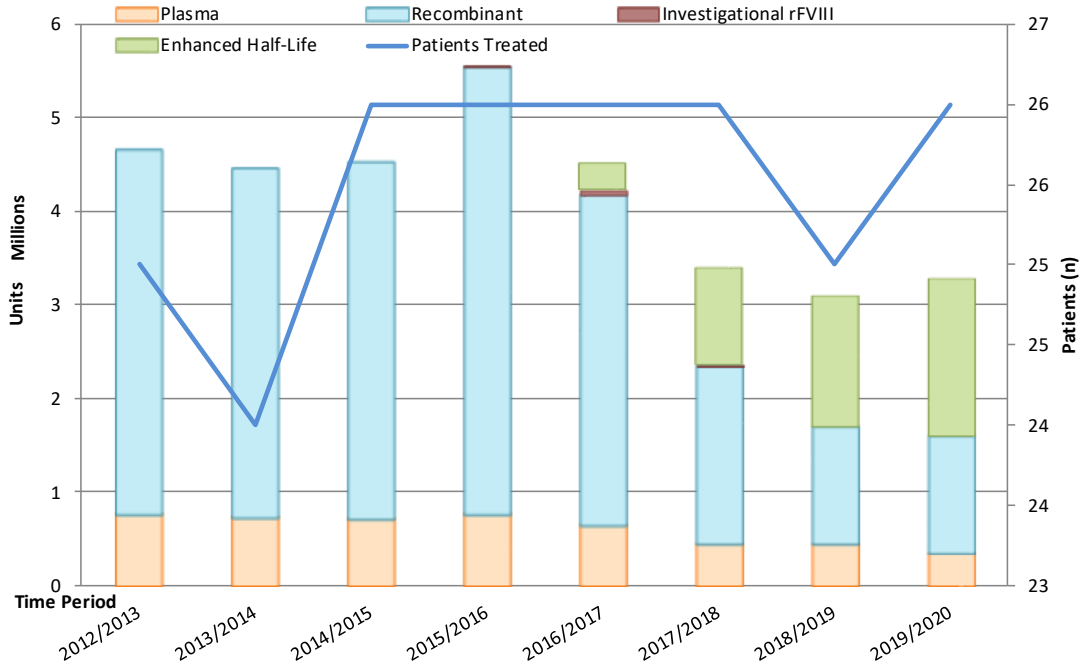


Figure 7b Factor IX units issued between April 2012 & March 2020 - *Severe Haemophilia B only*, people with a Scotland East postcode

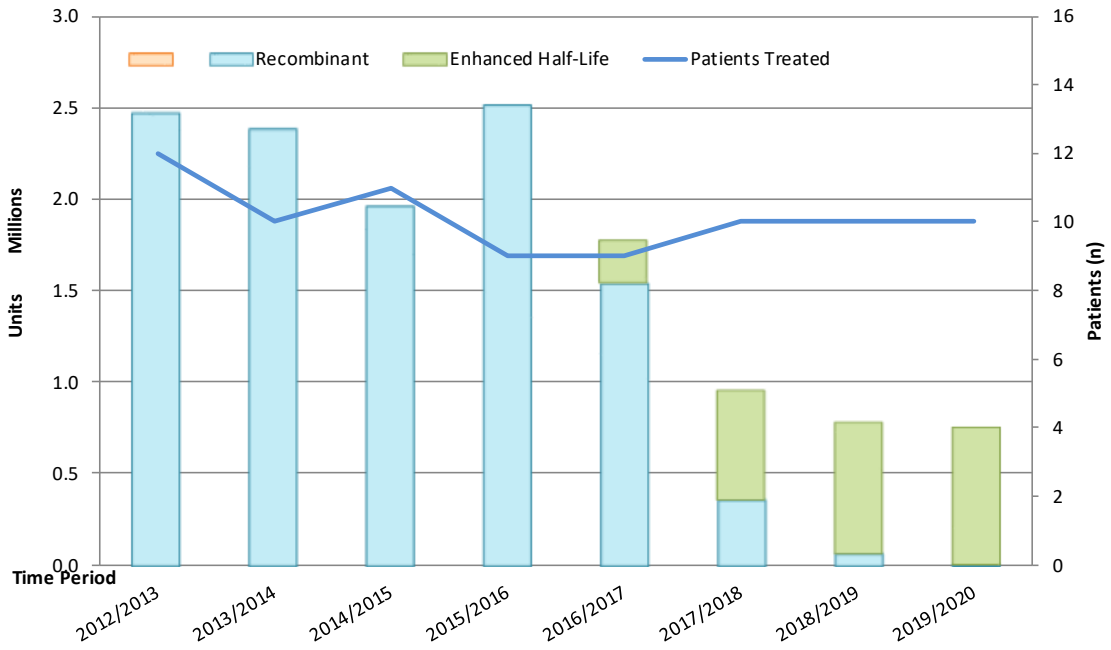
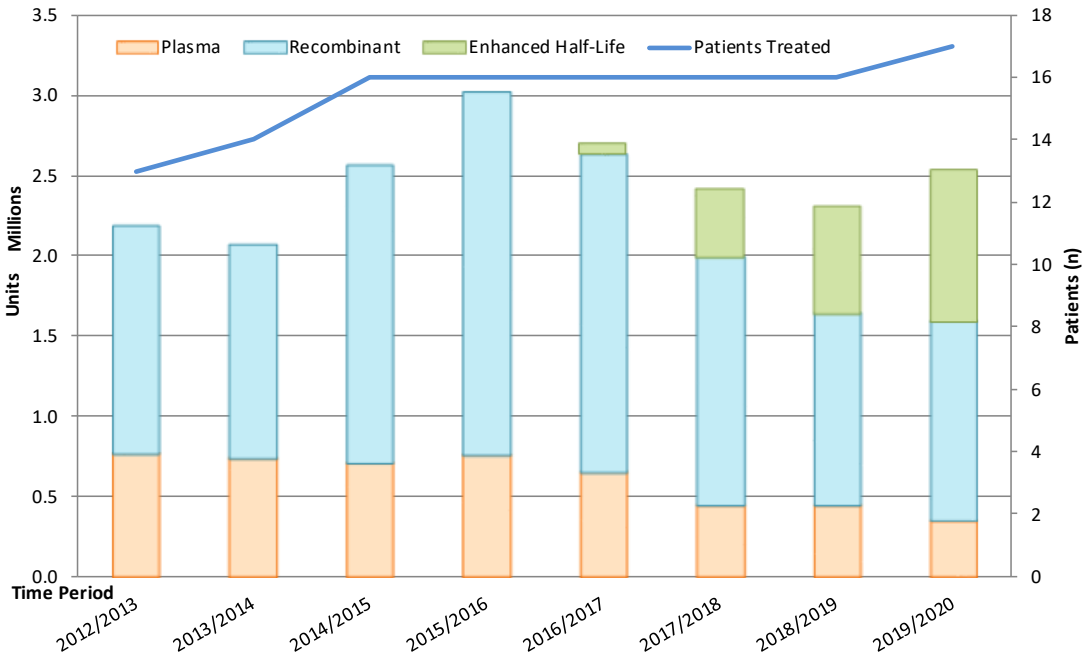


Figure 7c Factor IX units issued between April 2012 & March 2020 – *Severe Haemophilia B only*, people with a Scotland West postcode



Figures 7a to 7c give a historical view of the number of factor IX units issued between 2012/13 and 2019/20 for people with *severe haemophilia B only*. Figure 7a includes all people with a Scottish postcode. Figures 7b includes only people with a Scotland East postcode and Figure 7c those with a Scotland West postcode. The number of people treated is represented by the blue line using a secondary axis.

The data tables related to these graphs can be seen overleaf.

NOTE: Figure 7a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 7b and 7c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 7a is not directly comparable to Figures 7b and 7c as some people are dropped due to the fact that they were issued treatment from a haemophilia centre outside of Scotland. In addition, some people were issued treatment from both East and West Scotland.

Table 21a Data for Figure 7a - Factor IX units issued between April 2012 & March 2020 - *Severe Haemophilia B*, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2012/13	IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	759,000	1.00	3,876,983	1.00	0		0		4,635,983	1.00	25	1.00
2013/2014	728,000	0.96	3,712,792	0.96	0		0		4,440,792	0.96	24	0.96
2014/2015	699,000	0.92	3,810,000	0.98	0		0		4,509,000	0.97	26	1.04
2015/2016	748,500	0.99	4,763,250	1.23	2,500		0		5,514,250	1.19	26	1.04
2016/2017	646,500	0.85	3,512,000	0.91	47,653	1.00	293,750		4,499,903	0.97	26	1.04
2017/2018	442,500	0.58	1,899,000	0.49	25,588	0.54	1,019,500	1.00	3,386,588	0.73	26	1.04
2018/2019	441,000	0.58	1,261,750	0.33	0	0.00	1,380,250	1.35	3,083,000	0.67	25	1.00
2019/2020	340,000	0.45	1,252,250	0.32	0	0.00	1,680,750	1.65	3,273,000	0.71	26	1.04

Table 21b Data for Figure 7b - Factor IX units issued between April 2012 & March 2020 - *Severe Haemophilia B*, Scotland East postcodes

Year			Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
			IU	% difference since 2012/13	IU		IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013			2,461,733	1.00	0		0		2,461,733	1.00	12	1.00
2013/2014			2,375,542	0.96	0		0		2,375,542	0.96	10	0.83
2014/2015			1,953,000	0.79	0		0		1,953,000	0.79	11	0.92
2015/2016			2,502,000	1.02	0		0		2,502,000	1.02	9	0.75
2016/2017			1,536,000	0.62	0		230,500		1,766,500	0.72	9	0.75
2017/2018			360,500	0.15	0		585,750	1.00	946,250	0.38	10	0.83
2018/2019			66,000	0.03	0		716,500	1.22	782,500	0.32	10	0.83
2019/2020			6,000	0.00	0		746,500	1.27	752,500	0.31	10	0.83

Table 21c Data for Figure 7c - Factor IX units issued between April 2012 & March 2020 - *Severe Haemophilia B*, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2012/13	IU	% difference since 2012/13	IU		IU	% difference since 2017/18	IU	% difference since 2012/13	n	% difference since 2012/13
2012/2013	759,000	1.00	1,415,250	1.00	0		0		2,174,250	1.00	13	1.00
2013/2014	728,000	0.96	1,337,250	0.94	0		0		2,065,250	0.95	14	1.08
2014/2015	699,000	0.92	1,857,000	1.31	0		0		2,556,000	1.18	16	1.23
2015/2016	748,500	0.99	2,261,250	1.60	0		0		3,009,750	1.38	16	1.23
2016/2017	646,500	0.85	1,976,000	1.40	0		63,250		2,685,750	1.24	16	1.23
2017/2018	442,500	0.58	1,538,500	1.09	0		425,500	1.00	2,406,500	1.11	16	1.23
2018/2019	441,000	0.58	1,195,750	0.84	0		663,750	1.56	2,300,500	1.06	16	1.23
2019/2020	340,000	0.45	1,246,250	0.88	0		934,250	2.20	2,520,500	1.16	17	1.31

Von Willebrand Disease, Rarer Bleeding Disorders and Acquired Defects

Tables 22 - 24 shows the number of people with Scottish postcodes and reported products issued to treat von Willebrand disease, selected rarer disorders and acquired bleeding disorders during 2019/20, broken down by supplier.

Table 22 Concentrates issued to treat von Willebrand Disease

Manufacturer	Product	Patients Treated (n)	Total Units
CSL Behring	Voncento	66	810,000
Novo Nordisk	NovoSeven (mg)	1 - 2	176
	Desmopressin	52	1,237.7

Units in IU unless otherwise stated
Products containing VWF as well as FVIII are reported in FVIII units

Table 23 Concentrates issued to treat Rarer Bleeding Disorders

Manufacturer	Product	Patients Treated (n)	F.VII Deficiency	F.X Deficiency	F.XI Deficiency	F.XIII Deficiency
BPL	FXI	3	-	-	3,600	-
CSL Behring	Beriplex	1 - 2	-	6,000	-	-
	Fibrogammin P	4	-	-	-	83,750
Novo Nordisk	NovoSeven (mg)	7	14	-	-	-
	NovoThirteen	1 - 2	-	-	-	40,000

Units in IU unless otherwise stated

Table 24 Concentrates issued to treat Acquired Defects

Manufacturer	Product	Patients Treated (n)	Acquired Haemophilia A	Acquired von Willebrands
CSL Behring	Voncento	1 - 2	-	95,000
Novo Nordisk	NovoSeven (mg)	1 - 2	576	-
Takeda	Advate	1 - 2	68,000	-
	FEIBA	7	231,000	-
	OBIZUR	4	39,000	-
	Desmopressin	1 - 2	-	18

Products containing VWF as well as FVIII are reported in FVIII units

Table 25 Inhibitors by disease severity

Coagulation Defect	Severity (iu/dl) / Subtype	In Register *	Inhibitors		
			New n (%)	Ongoing n (%)	Historical n (%)
Haemophilia A	< 1	144	1 (0.7)	15 (10.4)	33 (22.9)
	1 - 5	69	0 (0.0)	2 (2.9)	4 (5.8)
	> 5	278	0 (0.0)	0 (0.0)	4 (1.4)
	Total	**491**	**1 (0.2)**	**17 (3.5)**	**41 (8.4)**
Haemophilia B	< 1	27	0 (0.0)	1 (3.7)	1 (3.7)
	1 - 5	43	0 (0.0)	0 (0.0)	0 (0.0)
	> 5	63	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**133**	**0 (0.0)**	**1 (0.8)**	**1 (0.8)**
von Willebrand disease	Type 3	10	0 (0.0)	1 (10.0)	1 (10.0)
	Others	1,133	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**1,185**	**0 (0.0)**	**1 (0.1)**	**1 (0.1)**

** Including patients not regularly treated*

Table 25 shows the incidence of new inhibitors during 2019/20, the prevalence of those still considered active and those considered inactive inhibitors for Haemophilia A, B and von Willebrand disease, broken down by disease severity.

Those labelled “new” were reported for the first time in the year 2019/20. Those labelled “ongoing” are those reported in previous years which have not been eradicated. Those reported as “historical” are those reported to have been previously eradicated or disappeared and not ongoing.

Table 26 Products issued to people with congenital bleeding disorders reported to have a positive inhibitor during 2019/20

Manufacturer	Product	Patients Treated (n)	Units
Haemophilia A			
Grifols	Fanhdi	1 - 2	97,000
Novo Nordisk	NovoEight	1 - 2	380,000
	NovoSeven (mg)	11	581
Pfizer	ReFacto AF	4	1,007,000
Roche	Hemlibra (mg)	10	48,180
Takeda	Advate	1 - 2	763,500
	FEIBA	3	1,161,000
CSL Behring	Desmopressin	1 - 2	55
	Investigational	1 - 2	720
Haemophilia B			
Novo Nordisk	NovoSeven (mg)	1 - 2	450
	Investigational	1 - 2	560
von Willebrand Disease			
Novo Nordisk	NovoSeven (mg)	1 - 2	176
Co-inherited diagnoses			
Novo Nordisk	NovoSeven (mg)	1 - 2	6
Roche	Hemlibra (mg)	1 - 2	6,300

Units in IU unless otherwise stated

Table 26 shows the number of people with Scottish postcodes and reported products issued to those with an inhibitor, newly reported or ongoing during 2019/20, broken down by diagnosis and supplier.

Table 27 Adverse Events

Adverse Event	Number of Patients	Number of Events
Allergic or Other Acute Event	0	0
Infection Event	0	0
Inhibitor Event	1	1
Intracranial Haemorrhage	1	1
Malignancy Event	8	9
Neurological Event	0	0
Other Event	0	0
Unexpected Poor Efficacy Event	0	0
Thrombosis Event	2	2
Total	**12**	**13**

See table 24 for breakdown of inhibitors by disease severity in Haemophilia A, B and von Willebrand disease

Table 27 shows the type and number of adverse events reported by Scottish centres during 2019/20.

Table 28 Causes of Death

Coagulation Defect	Cause of Death	Severity (factor level iu/dl)			
		< 1	1 - 5	> 5	Total
Haemophilia A	Carcinoma	0	0	2	**2**
	Cerebral haemorrhage	0	0	1	**1**
	Suicide	0	0	1	**1**
Haemophilia B	Cerebral haemorrhage	1	0	0	**1**
	Dementia/Alzheimer's disease	0	0	1	**1**
	Infection (Bacterial)	0	1	0	**1**
Acquired Haemophilia A	COAD	0	0	1	**1**
	Haemorrhage (misc)	0	1	0	**1**
	Infection (Bacterial)	0	0	2	**2**
	Ischaemic Heart Disease	1	0	0	**1**
Haemophilia A with Liver Transplant	Hepatocellular Carcinoma				**1**
von Willebrand disease	Carcinoma				**1**
	Infection (Bacterial)				**1**
	Unknown				**1**
Acquired von Willebrands	Infection (Bacterial)				**1**
F.VII deficiency	Unknown				**1**
F.XI Deficiency	Suicide				**1**
Hypodysfibrinogenemia	Infection (Bacterial)				**0**
	Total	**2**	**2**	**8**	**19**

Table 28 shows the causes of death reported in people with a Scottish postcode during 2019/20, broken down by diagnosis and disease severity.