



Bleeding Disorder Statistics for Scotland

April 2018 to March 2019

A report from the UK National Haemophilia Database

December 2019
Updated January 2020

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	1
Table 1 New registrations - Number of people newly registered between April 2018 & March 2019, by diagnosis and gender.....	1
Table 2 New registrations of haemophilia A & B between April 2018 & March 2019, by age and disease severity.....	2
In Register 3	
Table 3 In register – The total number of people in register as of 31 st March 2019 and the number treated between April 2018 & March 2019	3
Table 4 In Register – The total number of people in the register as of 31 st March 2019, by diagnosis and registered Haemophilia Centre.....	4
Table 5 In Register – The total number of people with Haemophilia A & B in the register as of 31 st March 2019, by severity and age group	5
Additional Figure Trend in people with Haemophilia A/B aged 60 years and above with Haemophilia A & B by age group.....	5
Table 6 In Register – The number of people with selected rarer bleeding disorders in the register as of 31 st March 2019 and the number treated between April 2018 & March 2019, by disease severity.....	7
Table 7 In register – The total number of people with Von Willebrand Disease in the register as of 31 st March 2019 and the number treated between April 2018 & March 2019, by disease severity, age group and gender	8
Treatment 9	
Table 8 People with a Scottish postcode, treated between April 2018 & March 2019 and region which issued the treatment, by diagnosis, all severities.....	9
Table 9 People with a non-Scottish postcode, registered & treated at a Scottish Haemophilia Centre between April 2018 & March 2019, by diagnosis, all severities.....	11
Haemophilia A and Factor VIII use	12
Table 10 Factor VIII issued, by diagnosis.....	12
Table 11 Products issued to treat Haemophilia A (including inhibitors).....	12
Table 12a Factor VIII issued to people with <i>Severe</i> Haemophilia A (incl. treatment for inhibitors), by issuing Haemophilia Centre.....	13
Table 12b Factor VIII issued by region for people with <i>Severe</i> Haemophilia A (incl. treatment for inhibitors), by region	13
Table 13 Factor VIII usage by Health Board for people with <i>Severe Haemophilia A</i> only (incl. treatment for inhibitors).....	14
Figure 1 Market share of factor VIII concentrates issued between April 2018 & March 2019	15
Figure 2a Factor VIII units issued between April 2011 & March 2019 – all diagnoses, all severities, all people with a Scottish postcode.....	16
Figure 2b Factor VIII units issued between April 2011 & March 2019 – all diagnoses, all severities, people with a Scotland East postcode	16

Figure 2c	Factor VIII units issued between April 2011 & March 2019 – all diagnoses, all severities, people with a Scotland West postcode.....	17
Table 14a	Data for Figure 2a - Factor VIII units issued between April 2011 & March 2019 – all diagnoses, all Scottish postcodes.....	18
Table 14b	Data for Figure 2b - Factor VIII units issued between April 2011 & March 2019 – all diagnoses, Scotland East postcodes.....	18
Table 14c	Data for Figure 2c - Factor VIII units issued between April 2011 & March 2019 – all diagnoses, Scotland West postcodes.....	19
Figure 3a	Factor VIII units issued between April 2011 & March 2019 – <i>Severe Haemophilia A only</i> , all people with a Scottish postcode	20
Figure 3b	Factor VIII units issued between April 2011 & March 2019 – <i>Severe Haemophilia A only</i> , people with a Scotland East postcode	20
Figure 3c	Factor VIII units issued between April 2011 & March 2019 – <i>Severe Haemophilia A only</i> , people with a Scotland West postcode.....	21
Table 15a	Data for Figure 3a - Factor VIII units issued between April 2011 & March 2019 – <i>Severe Haemophilia A</i> , all Scottish postcodes	22
Table 15b	Data for Figure 3b - Factor VIII units issued between April 2011 & March 2019 – <i>Severe Haemophilia A</i> , Scotland East postcodes	22
Table 15c	Data for Figure 3c - Factor VIII units issued between April 2011 & March 2019 – <i>Severe Haemophilia A</i> , Scotland West postcodes	23
Haemophilia B and Factor IX use		24
Table 16	Factor IX issued, by diagnosis	24
Table 17	Products issued to treat Haemophilia B (including inhibitors).....	24
Table 18a	Factor IX issued to people with <i>Severe Haemophilia B</i> (incl. treatment for inhibitors), by issuing Haemophilia Centre.....	25
Table 18b	Factor IX issued to patients with <i>Severe Haemophilia B</i> (incl. treatment for inhibitors), by region	25
Table 19	Factor IX usage by Health Board for people with <i>Severe Haemophilia B</i> only (incl. treatment for inhibitors).....	26
Figure 4	Market share of factor IX concentrates issued to people with a Scottish postcode between April 2018 & March 2019	27
Figure 5a	Factor IX units issued between April 2011 & March 2019 – all diagnoses, all severities, all people with a Scottish postcode.....	28
Figure 5b	Factor IX units issued between April 2011 & March 2019 – all diagnoses, all severities, people with a Scotland East postcode	28
Figure 5c	Factor IX units issued between April 2011 & March 2019 – all diagnoses, all severities, people with a Scotland West postcode.....	29
Table 20a	Data for figure 5 - Factor IX units issued between April 2011 & March 2019 – all diagnoses, all Scottish postcodes	30
Table 20b	Data for figure 5b - Factor IX units issued between April 2011 & March 2019 – all diagnoses, Scotland East postcodes.....	30
Table 20c	Data for Figure 5c - Factor IX units issued between April 2011 & March 2019 – all diagnoses, Scotland West postcodes.....	31
Figure 6a	Factor IX units issued between April 2011 & March 2019 – <i>Severe Haemophilia B only</i> , all people with a Scottish postcode	32

Figure 6b	Factor IX units issued between April 2011 & March 2019 – <i>Severe Haemophilia B only</i> , people with a Scotland East postcode	32
Figure 6c	Factor IX units issued between April 2011 & March 2019 – <i>Severe Haemophilia B only</i> , people with a Scotland West postcode	33
Table 21a	Data for Figure 6a - Factor IX units issued between April 2011 & March 2019 – <i>Severe Haemophilia B</i> , all Scottish postcodes	34
Table 21b	Data for Figure 6b - Factor IX units issued between April 2011 & March 2019 – <i>Severe Haemophilia B</i> , Scotland East postcodes	34
Table 21c	Data for Figure 6c - Factor IX units issued between April 2011 & March 2019 – <i>Severe Haemophilia B</i> , Scotland West postcodes	35
Von Willebrand Disease, Rarer Bleeding Disorders and Acquired Defects		36
Table 21	Concentrates issued to treat von Willebrand Disease	36
Table 22	Concentrates issued to treat Rarer Bleeding Disorders	36
Table 23	Concentrates issued to treat Acquired Defects.....	36
Adverse Events and Deaths.....		37
Table 24	Inhibitors by disease severity	37
Table 25	Products issued to people with congenital bleeding disorders reported to have a positive inhibitor during 2018/19.....	38
Table 26	Adverse Events	39
Table 27	Causes of Death	40

New Registrations

Table 1 New registrations - Number of people newly registered between April 2018 & March 2019, by diagnosis and gender

Coagulation Defect	Male	Female	Total
Haemophilia A	22	0	22
Haemophilia A Carrier		6	6
Acquired Haemophilia A	7	4	11
Haemophilia A with Liver Transplant	1	0	1
Haemophilia B	5	0	5
Haemophilia B Carrier		3	3
von Willebrand disease	15	17	32
Probable von Willebrands disease	0	1	1
Acquired von Willebrands	0	2	2
F.V deficiency	2	5	7
F.VII deficiency	17	21	38
F.X deficiency	1	1	2
F.XI Deficiency	13	11	24
F.XIII Deficiency	0	1	1
Dysfibrinogenemia	8	9	17
Hypofibrinogenemia	2	2	4
Platelet defects	6	15	21
Miscellaneous	0	6	6
Unclassified	0	11	11
Total	99	115	214

*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 2018 & March 2019, 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	1	4	7
	10 : 19	0	1	0	1
	20 : 29	0	0	6	6
	30 : 39	0	0	1	1
	40 : 49	0	0	1	1
	50 : 59	0	0	1	1
	60 : 69	0	0	4	4
	70 +	0	0	1	1
Total		2	2	18	22
Haemophilia B	0 : 9	0	0	3	3
	10 : 19	0	0	1	1
	20 : 29	0	0	1	1
	30 : 39	0	0	0	0
	40 : 49	0	0	0	0
	50 : 59	0	0	0	0
	60 : 69	0	0	0	0
	70 +	0	0	0	0
Total		0	0	5	5

N.B Age calculated at mid-year, 30/09/2018

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 2019 and the number treated between April 2018 & March 2019

Coagulation Defect	In Register			Treated (n)	Treated %
	Males	Females	Total		
Haemophilia A	485	1	486	287	59.05%
Acquired Haemophilia A	18	24	42	4	9.52%
Haemophilia A Carrier		242	242	14	5.79%
Haemophilia A with Liver Transplant	4	0	4	1	25.00%
Haemophilia B	127	0	127	67	52.76%
Haemophilia B Carrier		69	69	4	5.80%
Haemophilia B with Liver Transplant	1	0	1	0	0.00%
von Willebrand disease	394	764	1,158	93	8.03%
Acquired von Willebrand Disease	8	14	22	3	13.64%
Probable von Willebrands disease	15	43	58	2	3.45%
Prothrombin Deficiency	1	2	3	1	33.33%
F.V deficiency	9	19	28	0	0.00%
F.VII deficiency	105	129	234	8	3.42%
F.X deficiency	15	30	45	1	2.22%
F.XI Deficiency	95	124	219	4	1.83%
F.XIII Deficiency	2	3	5	4	80.00%
Combined V+VIII Deficiency	1	2	3	2	66.67%
Co-inherited diagnoses	6	13	19	1	5.26%
Acquired F.V deficiency	0	1	1	0	0.00%
Acquired F.XIII Deficiency	0	1	1	0	0.00%
Acquired Deficiency (other)	1	0	1	0	0.00%
Fibrinogen Deficiency	0	3	3	0	0.00%
Dysfibrinogenemia	83	132	215	1	0.47%
Hypofibrinogenemia	10	14	24	0	0.00%
Hypodysfibrinogenemia	7	5	12	1	8.33%
Glanzmanns Thrombasthenia	3	9	12	3	25.00%
Bernard Soulier	3	4	7	0	0.00%
Platelet Defect	57	152	209	7	3.35%
Miscellaneous	8	29	37	0	0.00%
Unclassified	5	52	57	0	0.00%
Totals	1,463	1,881	3,344	508	

Table 3 shows the total number of active registrations of people with a Scottish postcode and the number who were issued treatment during 2018/19.

Table 4 In Register - The total number of people in the register as of 31st March 2019, by diagnosis and registered Haemophilia Centre

Coagulation Defect	Aberdeen	Dundee	Edinburgh	Glasgow	Inverness	Total
Haemophilia A	72	48	125	246	31	522
Haemophilia A Carrier	29	23	33	155	6	246
Haemophilia A with Liver Transplant	1	1	2	0	0	4
Acquired Haemophilia A	6	2	21	16	0	45
Haemophilia B	8	12	30	80	2	132
Haemophilia B Carrier	1	13	14	43	0	71
Haemophilia B with Liver Transplant	0	1	0	0	0	1
von Willebrand disease	165	185	160	655	41	1206
Acquired von Willebrands	2	3	11	10	0	26
Probable von Willebrands disease	1	35	7	17	0	60
F.V deficiency	6	0	3	19	0	28
F.VII deficiency	41	57	28	111	1	238
F.X deficiency	9	5	6	25	0	45
F.XI Deficiency	21	46	55	99	4	225
F.XIII Deficiency	2	0	1	2	0	5
Combined V+VIII Deficiency	0	0	1	2	0	3
Co-inherited diagnoses	0	6	2	11	0	19
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
Dysfibrinogenemia	3	20	36	161	1	221
Hypofibrinogenemia	4	0	3	17	2	26
Hypodysfibrinogenemia	0	0	11	1	0	12
Glanzmanns Thrombasthenia	1	3	5	3	0	12
Bernard Soulier	1	1	2	4	0	8
Platelet Defect	4	17	98	91	0	210
Miscellaneous	2	9	2	24	0	37
Unclassified	0	1	3	53	0	57
Total	380	488	661	1848	88	3,465

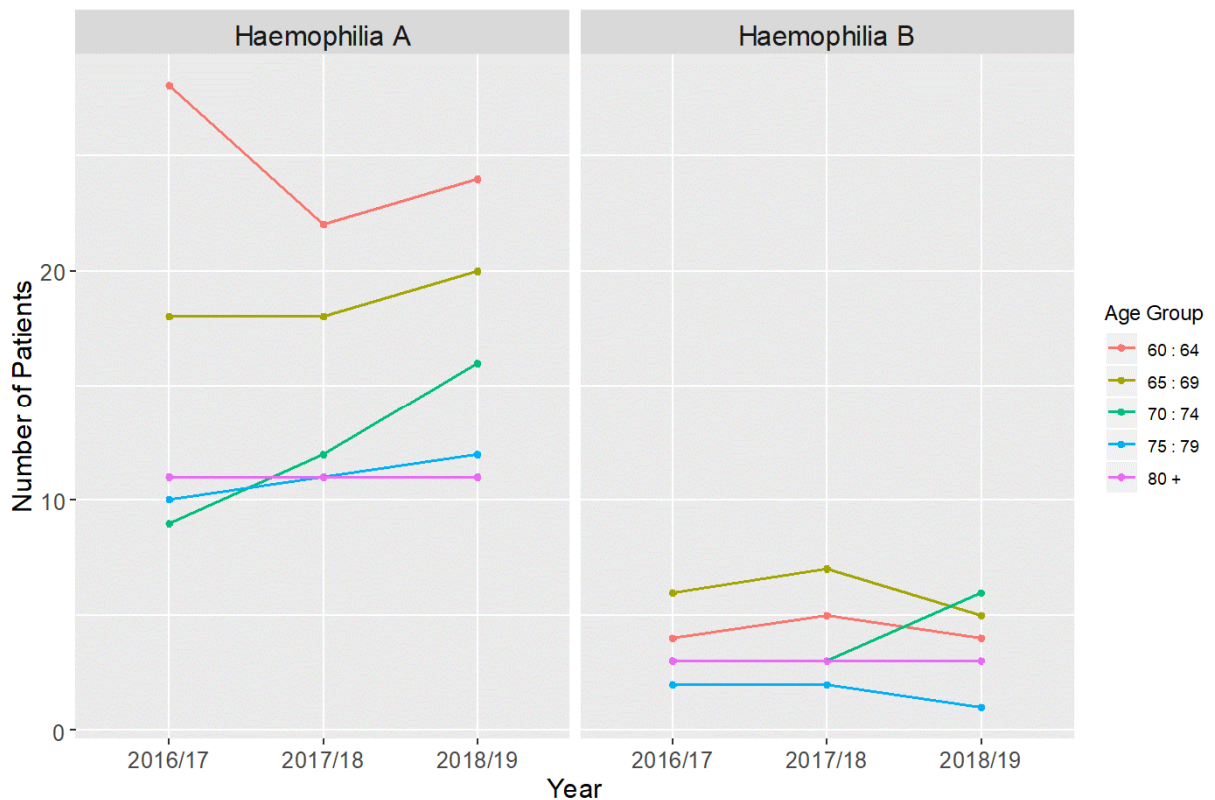
Table 4 shows the number of people registered at each Haemophilia Centre. Patients are allocated to their registered haemophilia centre regardless of their home postcode.

Table 5 In Register - The total number of people with Haemophilia A & B in the register as of 31st March 2019, 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	50	15	55	120
	≥18 years	96	55	215	366
Total		146	70	270	486
Haemophilia B	<18 years	7	7	13	27
	≥18 years	18	36	46	100
Total		25	43	59	127

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.

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



Data tables for additional figure

		Haemophilia A			
Year	Age (years)	Number of Patients (factor VIII level iu/dl)			Total
		< 1	1 - 5	> 5	
2016/17	60 : 64	1	6	21	28
	65 : 69	4	3	11	18
	70 : 74	1	1	7	9
	75 : 79	1	1	8	10
	80 +	0	1	10	11
2017/18	60 : 64	2	4	16	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	4	17	24
	65 : 69	3	2	15	20
	70 : 74	2	2	12	16
	75 : 79	1	2	9	12
	80 +	0	1	10	11

		Haemophilia B			
Year	Age (years)	Number of Patients (factor IX level iu/dl)			Total
		< 1	1 - 5	> 5	
2016/17	60 : 64	1	0	3	4
	65 : 69	0	2	4	6
	70 : 74	0	2	1	3
	75 : 79	1	0	1	2
	80 +	0	1	2	3
2017/18	60 : 64	1	0	4	5
	65 : 69	0	2	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	0	2	3	5
	70 : 74	0	2	4	6
	75 : 79	1	0	0	1
	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 2019 and the number treated between April 2018 & March 2019, 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	27	0	0	0	28	0
F.VII deficiency	7	1	226	7	1	0	234	8
F.X deficiency	0	0	45	1	0	0	45	1
F.XI Deficiency	9	1	209	3	1	0	219	4
Total	17	2	507	11	2	-	526	13

Coagulation Defect	<2		2 - <10		10 - <15		Total	
	In Reg	Treated	In Reg	Treated	In Reg	Treated	In Reg	Treated
F.XIII Deficiency	2	2	2	2	1	0	5	4
Total	2	2	2	2	1	-	5	4

Table 6 shows the number of people with selected rarer bleeding disorders and a Scottish postcode known to the NHD during 2018/19. 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 2019 and the number treated between April 2018 & March 2019, 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	0	13	11	0	24	1	40	70	0	111	135	10
Type 2A	1	0	0	0	1	1	5	0	0	6	7	1
Type 2B	0	0	0	0	0	0	2	1	0	3	3	0
Type 2M	0	2	0	0	2	1	2	2	0	5	7	0
Type 2N	0	0	0	0	0	0	0	1	0	1	1	0
Type 2 Unspecified	1	0	0	0	1	1	1	5	0	7	8	3
Type 3	1				1	5				5	6	5
Type Unreported	6	21	28	0	55	12	48	105	0	165	220	13
Low VWF	0	0	2	0	2	0	0	5	0	5	7	1
Sub Total Males											394	33
Females												
Type 1	1	9	8	0	18	3	76	179	0	258	276	15
Type 2A	1	0	0	0	1	7	8	1	0	16	17	5
Type 2B	0	0	1	0	1	0	5	4	0	9	10	1
Type 2M	1	2	0	0	3	7	13	0	0	20	23	2
Type 2N	0	0	0	0	0	1	2	1	0	4	4	1
Type 2 Unspecified	1	1	0	0	2	0	6	2	0	8	10	0
Type 3	1				1	3				3	4	1
Type Unreported	8	19	21	0	48	18	84	256	1	359	407	33
Low VWF	0	1	0	0	1	0	0	12	0	12	13	2
Sub Total Females											764	60
Grand Total - Males and Females											1,158	93

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 2018 & March 2019 and region which issued the treatment, by diagnosis, all severities

Coagulation Defect	Region Issuing Treatment	Patients Treated (n)
Haemophilia A	Cheshire, Warrington & Wirral	1
	Cumbria, Northumberland & Tyne and Wear	1
	South Yorkshire & Bassetlaw	1
	Scotland East	139
	Scotland West	149
Sub total		291
Haemophilia A Carrier	East Anglia	1
	Scotland East	5
	Scotland West	9
Sub total		15
Acquired Haemophilia A	Scotland East	3
	Scotland West	1
Sub total		4
Haemophilia A with Liver Transplant	Scotland East	1
Sub total		1
Haemophilia B	London	1
	Scotland East	23
	Scotland West	46
Sub total		70
Haemophilia B Carrier	Scotland West	4
Sub total		4
von Willebrand disease	Birmingham & Black Country	1
	Scotland East	49
	Scotland West	43
Sub total		93
Probable von Willebrand disease	Scotland East	2
Sub total		2
Acquired von Willebrand Disease	Scotland East	3
Sub total		3
F.VII deficiency	Scotland East	3
	Scotland West	5
Sub total		8
F.X deficiency	Scotland West	1
Sub total		1
F.XI Deficiency	Scotland East	2
	Scotland West	2
Sub total		4
F.XIII Deficiency	Scotland East	3
	Scotland West	1
Sub total		4

Continued overleaf...

Table 8 continued...

Coagulation Defect	Region	Patients Treated (n)
Combined V+VIII Deficiency	Scotland East	1
	Scotland West	1
Sub total		2
Co-inherited diagnoses	Scotland East	1
Sub total		1
Dysfibrinogenemia	Scotland West	1
Sub total		1
Hypodysfibrinogenemia	Scotland East	1
Sub total		1
Prothrombin Deficiency	Scotland West	1
Sub total		1
Glanzmann's Thrombasthenia	Scotland East	2
	Scotland West	1
Sub total		3
Other platelet defects	Scotland East	4
	Scotland West	3
Sub total		7
Grand total		516

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 patients 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 2018 & March 2019, by diagnosis, all severities

Coagulation Defect	Patient's home postcode region	Patients Registered (n)	Patients Treated (n)
Haemophilia A	London	6	3
	Midlands and East of England	3	3
	North of England	5	5
	South East	7	3
	South West	3	3
Sub total		24	17
Haemophilia A Carrier	Midlands and East of England	2	-
	North of England	1	-
Sub total		3	-
Haemophilia B	Midlands and East of England	1	-
	North of England	1	1
Sub total		2	1
von Willebrand disease	London	2	1
	Midlands and East of England	1	1
	North of England	14	1
	Northern Ireland	1	-
	South East	2	1
Sub total		20	4
Acquired von Willebrands	South East	2	1
Sub total		2	1
F.VII deficiency	North of England	2	-
	South East	1	-
Sub total		3	-
F.XI Deficiency	North of England	1	-
	South East	1	-
	South West	1	-
Sub total		3	-
Dysfibrinogenemia	Midlands and East of England	1	-
Hypofibrinogenemia	London	1	-
Grand total		59	23

The patients reported in Table 9 were registered at or issued treatment from a Scottish Haemophilia Centre during 2018/19, 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	FVIII (IU)				Total
		Plasma	Recombinant	Enhanced Half-Life	Investigational	
Haemophilia A	266	3,340,000	44,275,750	4,580,950	-	52,196,700
Haemophilia A Carrier	11	-	79,500	-	-	79,500
Haemophilia A with Liver Transplant	1	-	62,500	-	-	62,500
von Willebrand disease	51	712,000	-	-	-	712,000
Acquired von Willebrands	3	24,500	-	-	-	24,500
Total	332	4,076,500	44,417,750	4,580,950	-	53,075,200

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 2018/19. Also shown are the number of units issued, broken down by diagnosis and product type.

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

Manufacturer	Product	Total Units
Bayer	Kogenate	418,000
CSL Behring	Helixate Nexgen	34,000
Grifols	Fanhdi	3,340,000
Novo Nordisk	NovoEight	7,795,500
	NovoSeven (mg)	684
Octapharma	Nuwiq	90,000
Pfizer	ReFacto AF	17,505,750
Roche	Hemlibra (mg)	22,463
SOBI/Biogen	Elocta	4,580,950
Takeda	Advate	18,432,500
	FEIBA	3,121,000

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	5,546,750	264,131
Dundee	16	4,222,750	263,922
Edinburgh	29	9,608,000	331,310
Glasgow	68	18,961,450	278,845
Inverness	3	335,000	111,667
	137	38,673,950	282,292

Table 12a reports the number of people with severe haemophilia A treated and the number of units of factor VIII issued during 2018/19. This is broken down by the haemophilia centre which issued the treatment.

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	69	19,712,500	285,688
Scotland West	68	18,961,450	278,845
	137	38,673,950	282,292

Table 12b reports the number of people with severe haemophilia A treated and the number of units of factor VIII issued during 2018/19. This is broken down by region based on the person's postcode as recorded on the NHD.

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	115,270	2	1,881,000	940,500	16.32
Dumfries and Galloway	148,790	3	1,532,000	510,667	10.30
Western Isles	26,830	1	487,500	487,500	18.17
Forth Valley	306,070	6	2,878,500	479,750	9.40
Lothian	897,770	22	6,776,000	308,000	7.55
Grampian	584,550	19	5,780,000	304,211	9.89
Tayside	416,080	11	3,018,750	274,432	7.26
Lanarkshire	659,200	6	1,574,500	262,417	2.39
Ayrshire and Arran	369,670	11	2,605,000	236,818	7.05
Greater Glasgow and Clyde	1,174,980	37	8,695,950	235,026	7.40
Highland	321,800	6	1,389,000	231,500	4.32
Shetland	22,990	3	570,750	190,250	24.83
Fife	371,910	8	1,485,000	185,625	3.99
Scotland	5,438,100	135	38,673,950	286,474	7.11

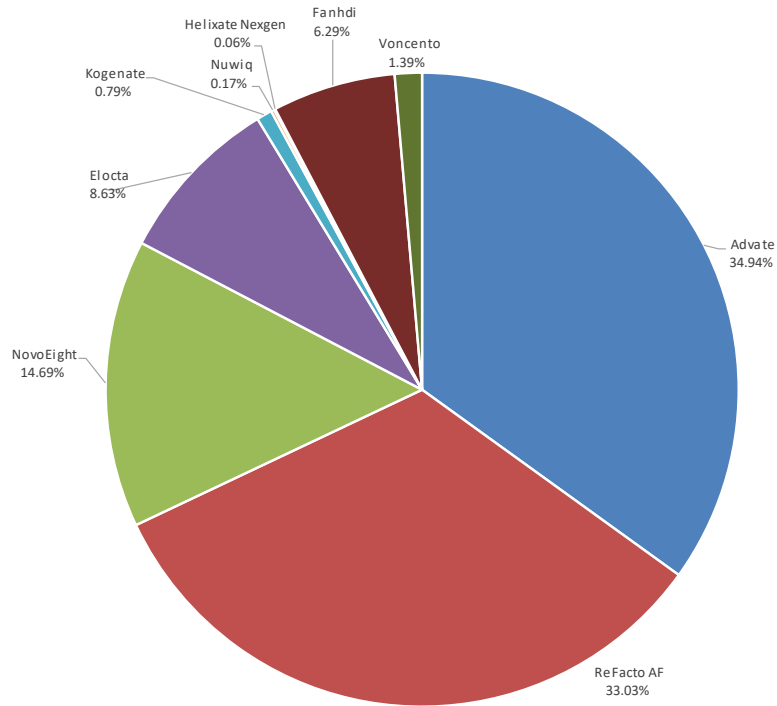
Ranked by mean usage

Mid-2018 population estimates Scotland supplied by National Records of Scotland under the Open Government Licence v3.0 © Crown Copyright 2019

Table 13 reports the number of people with severe haemophilia A treated 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 duplicate patients. People are allocated to a Health Board based on their home postcode.

Figure 1 Market share of factor VIII concentrates issued between April 2018 & March 2019



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 (n)
Takeda	Advate	18,543,000	108
Pfizer	ReFacto AF	17,532,250	121
Novo Nordisk	NovoEight	7,798,500	45
SOBI/Biogen	Elocta	4,580,950	19
Bayer	Kogenate	420,000	6
Octapharma	Nuwiq	90,000	1
CSL Behring	Helixate Nexgen	34,000	1
Grifols	Fanhdi	3,340,000	3
CSL Behring	Voncento	736,500	54
	Total	53,075,200	358

Figure 1 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 patients treated with more than one product type. The patient numbers in Table 10 do not contain duplicates.

Figure 2a Factor VIII units issued between April 2011 & March 2019 - all diagnoses, all severities, all people with a Scottish postcode

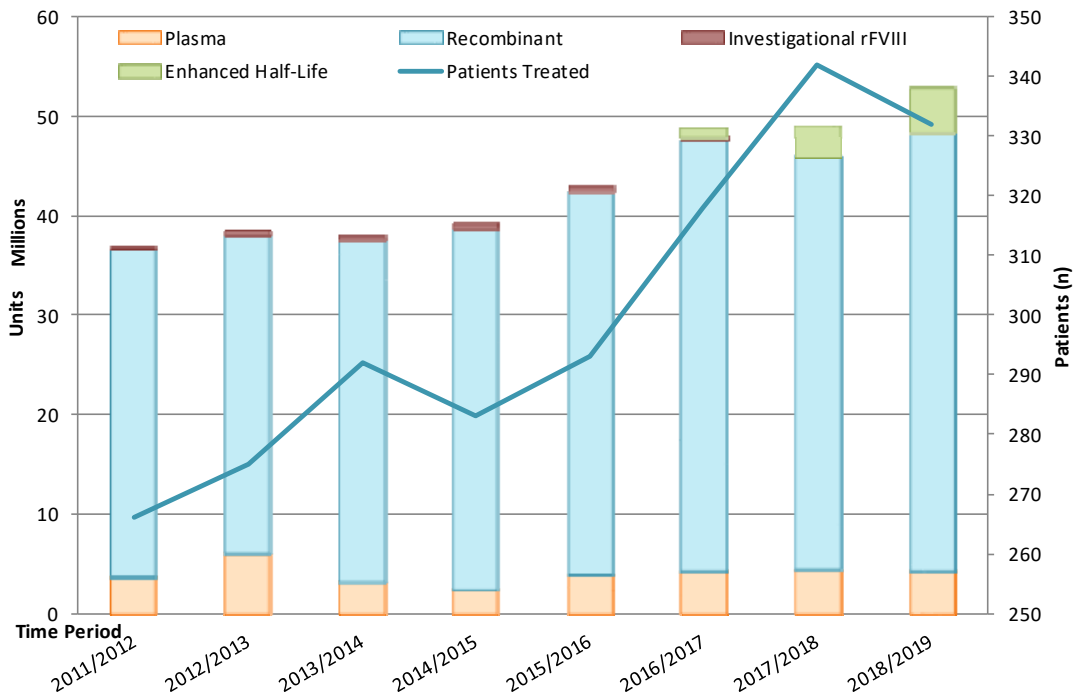


Figure 2b Factor VIII units issued between April 2011 & March 2019 - all diagnoses, all severities, people with a Scotland East postcode

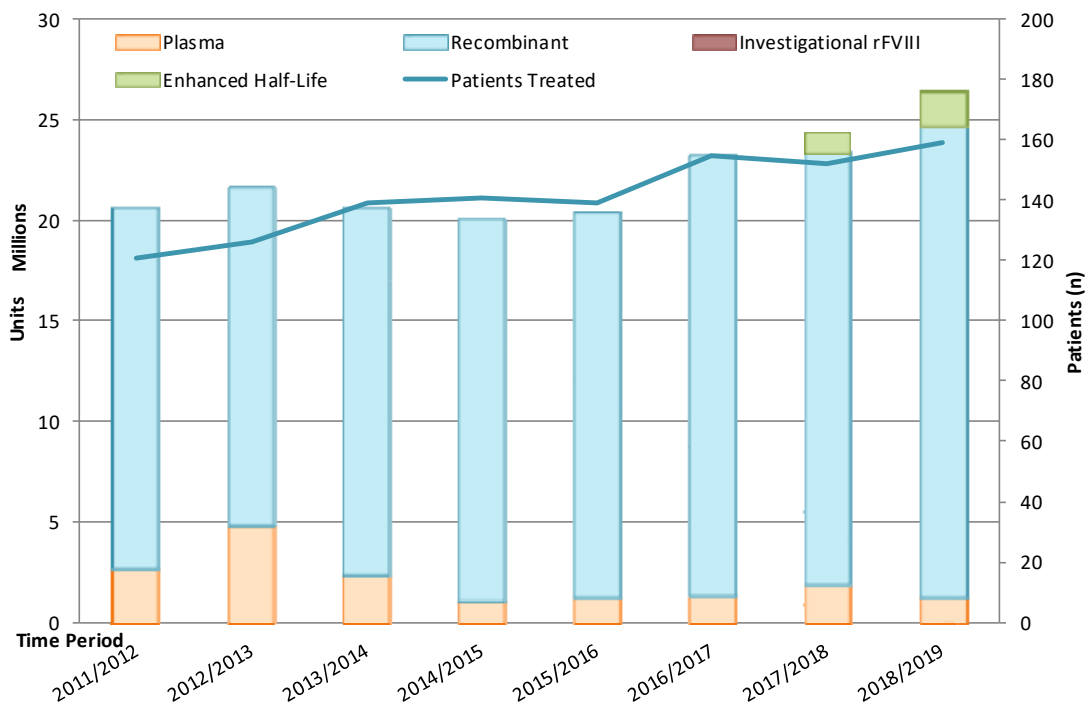
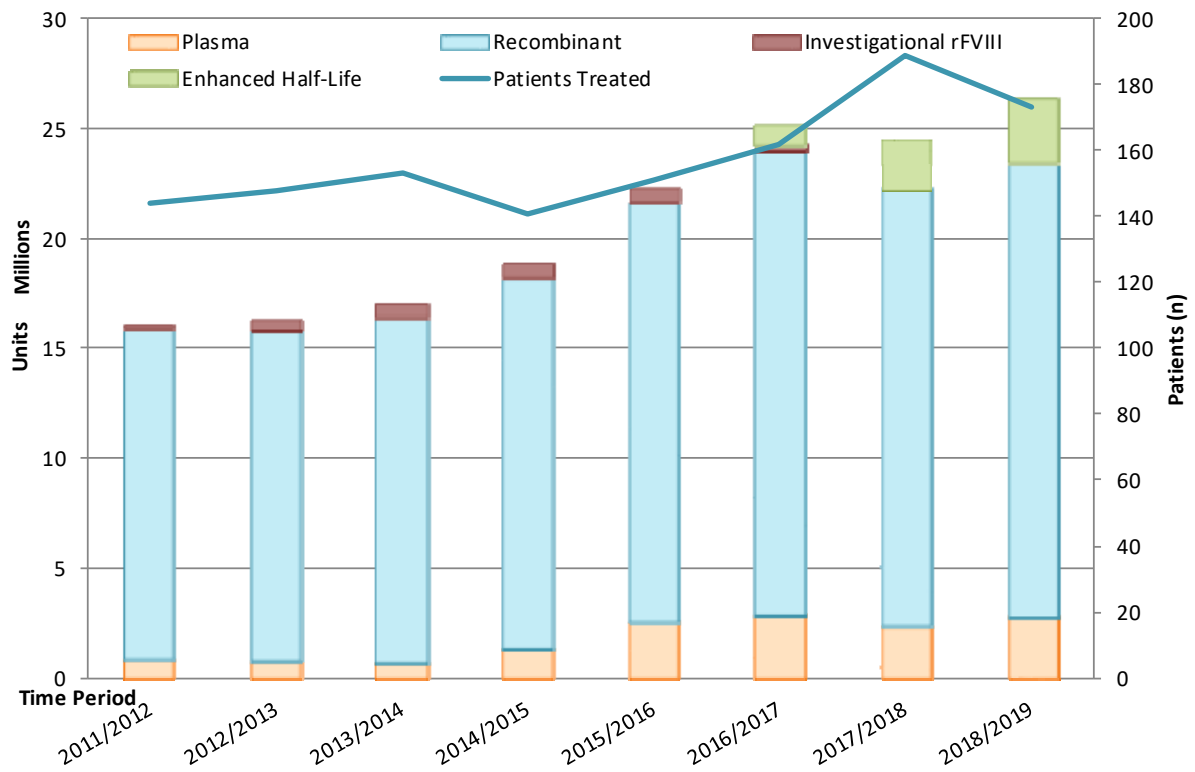


Figure 2c Factor VIII units issued between April 2011 & March 2019 - all diagnoses, all severities, people with a Scotland West postcode



Figures 2a to 2c include products containing a combination of VWF and FVIII, which are reported in FVIII units

Figures 2a to 2c give an historical view of the number of factor VIII units issued between 2011/12 and 2018/19 for all diagnoses and all severities. Figure 2a includes all people with a Scottish postcode. Figures 2b only includes people with a Scotland East postcode and Figure 2c 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 2a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 2b and 2c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 2a is not directly comparable to Figures 2b and 2c 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 2a - Factor VIII units issued between April 2011 & March 2019 - all diagnoses, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	3,588,650	1.00	33,361,259	1.00	137,500	1.00	0		37,087,409	1.00	266	1.00
2012/2013	5,866,400	1.63	32,352,475	0.97	472,202	3.43	0		38,691,077	1.04	275	1.03
2013/2014	3,148,100	0.88	34,468,895	1.03	630,000	4.58	0		38,246,995	1.03	292	1.10
2014/2015	2,585,000	0.72	36,263,500	1.09	674,750	4.91	0		39,523,250	1.07	283	1.06
2015/2016	3,761,050	1.05	38,836,250	1.16	613,500	4.46	0		43,210,800	1.17	293	1.10
2016/2017	4,167,000	1.16	43,602,750	1.31	330,500	2.40	849,750	1.00	48,950,000	1.32	318	1.20
2017/2018	4,228,000	1.18	41,858,250	1.25	0	0.00	3,089,750	3.64	49,176,000	1.33	342	1.29
2018/2019	4,076,500	1.14	44,417,750	1.33	0	0.00	4,580,950	5.39	53,075,200	1.43	332	1.25

Table 14b Data for Figure 2b - Factor VIII units issued between April 2011 & March 2019 - all diagnoses, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU		IU	% difference since 2017/18	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	2,642,200	1.00	17,984,758	1.00	0		0		20,626,958	1.00	121	1.00
2012/2013	4,846,900	1.83	16,835,225	0.94	0		0		21,682,125	1.05	126	1.04
2013/2014	2,298,100	0.87	18,347,645	1.02	0		0		20,645,745	1.00	139	1.15
2014/2015	1,041,000	0.39	19,003,750	1.06	0		0		20,044,750	0.97	141	1.17
2015/2016	1,167,000	0.44	19,237,500	1.07	0		0		20,404,500	0.99	139	1.15
2016/2017	1,258,000	0.48	22,003,500	1.22	0		0		23,261,500	1.13	155	1.28
2017/2018	1,795,500	0.68	21,680,250	1.21	0		919,000	1.00	24,394,750	1.18	152	1.26
2018/2019	1,186,000	0.45	23,573,750	1.31	0		1,720,000	1.87	26,479,750	1.28	159	1.31

Table 14c Data for Figure 2c - Factor VIII units issued between April 2011 & March 2019 - all diagnoses, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	856,450	1.00	15,060,501	1.00	137,500	1.00	0		16,054,451	1.00	144	1.00
2012/2013	771,500	0.90	15,064,750	1.00	472,202	3.43	0		16,308,452	1.02	148	1.03
2013/2014	694,000	0.81	15,692,750	1.04	630,000	4.58	0		17,016,750	1.06	153	1.06
2014/2015	1,383,000	1.61	16,775,250	1.11	674,750	4.91	0		18,833,000	1.17	141	0.98
2015/2016	2,470,550	2.88	19,246,750	1.28	613,500	4.46	0		22,330,800	1.39	151	1.05
2016/2017	2,771,000	3.24	21,179,250	1.41	330,500	2.40	849,750	1.00	25,130,500	1.57	162	1.13
2017/2018	2,316,500	2.70	20,024,500	1.33	0	0.00	2,170,750	2.55	24,511,750	1.53	189	1.31
2018/2019	2,746,500	3.21	20,778,500	1.38	0	0.00	2,860,950	3.37	26,385,950	1.64	173	1.20

Figure 3a Factor VIII units issued between April 2011 & March 2019 - *Severe Haemophilia A only*, all people with a Scottish postcode

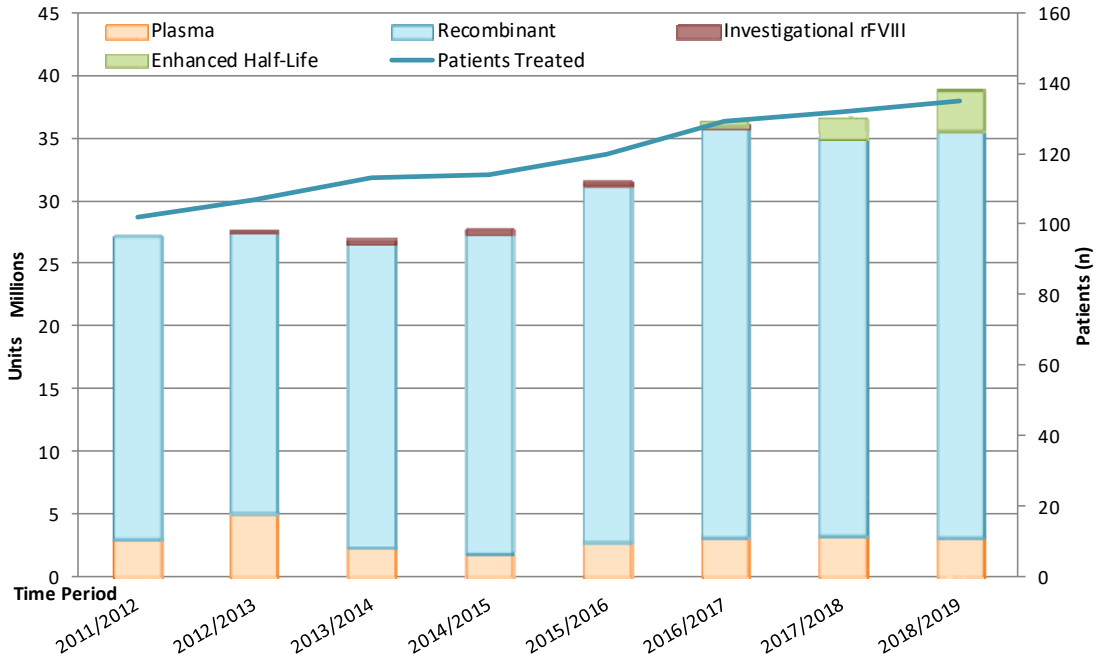


Figure 3b Factor VIII units issued between April 2011 & March 2019 - *Severe Haemophilia A only*, people with a Scotland East postcode

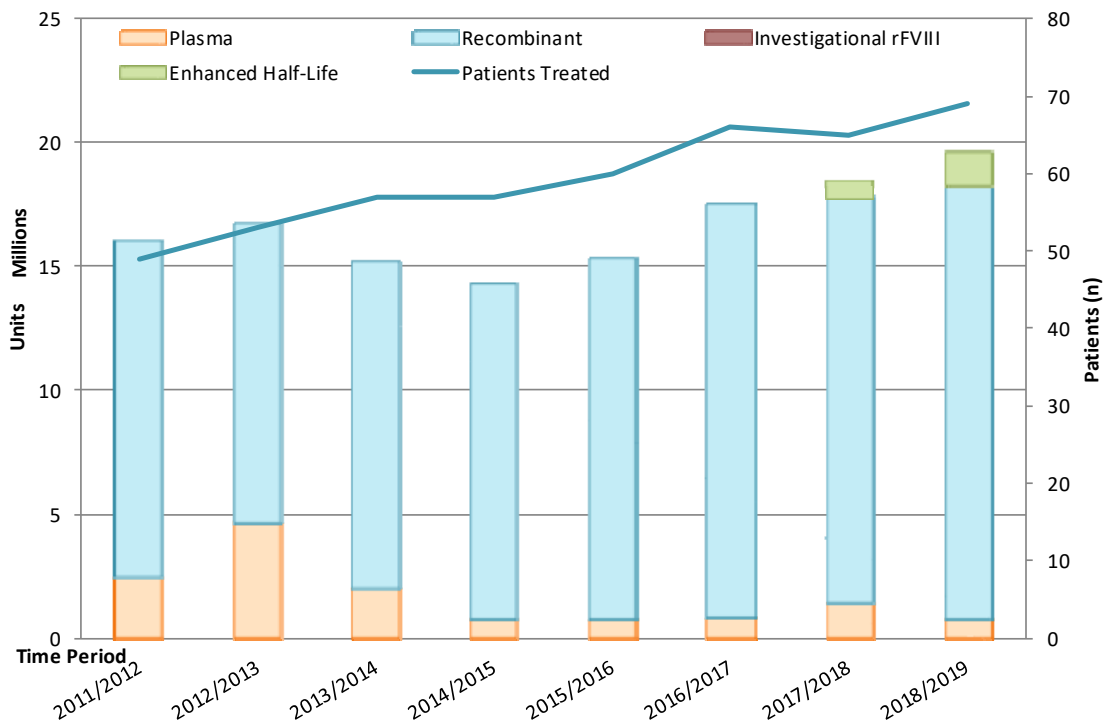
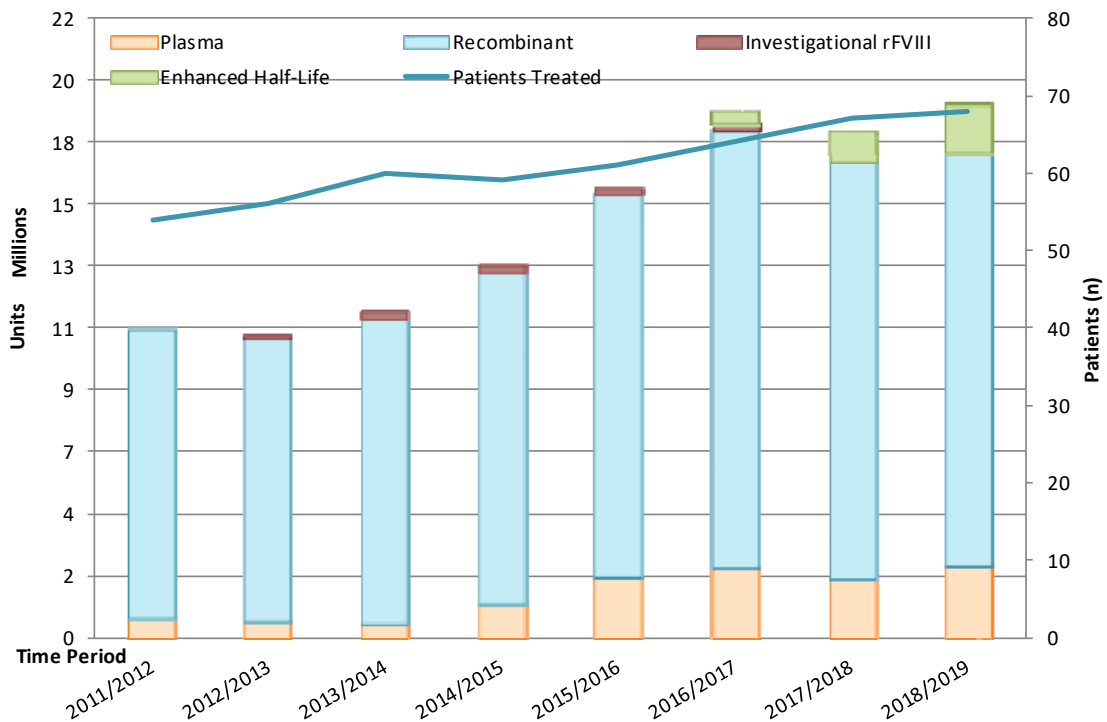


Figure 3c Factor VIII units issued between April 2011 & March 2019 - *Severe Haemophilia A only*, people with a Scotland West postcode



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

Figures 3a to 3c give an historical view of the number of factor VIII units issued between 2011/12 and 2018/19 for people with *severe haemophilia A only*. Figure 3a includes all patients with a Scottish postcode. Figure 3b includes only 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 15a Data for Figure 3a - Factor VIII units issued between April 2011 & March 2019 - *Severe Haemophilia A*, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2012/13	IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	3,100,000	1.00	23,991,166	1.00	0		0		27,091,166	1.00	102	1.00
2012/2013	5,143,500	1.66	22,309,988	0.93	105,202	1.00	0		27,558,690	1.02	107	1.05
2013/2014	2,456,000	0.79	24,128,855	1.01	293,000	2.79	0		26,877,855	0.99	113	1.11
2014/2015	1,923,000	0.62	25,432,250	1.06	314,750	2.99	0		27,670,000	1.02	114	1.12
2015/2016	2,872,500	0.93	28,274,250	1.18	253,500	2.41	0		31,400,250	1.16	120	1.18
2016/2017	3,295,500	1.06	32,354,500	1.35	240,500	2.29	430,750	1.00	36,321,250	1.34	129	1.26
2017/2018	3,459,000	1.12	31,386,500	1.31	0	0.00	1,716,250	3.98	36,561,750	1.35	132	1.29
2018/2019	3,340,000	1.08	32,224,750	1.34	0	0.00	3,109,200	7.22	38,673,950	1.43	135	1.32

Table 15b Data for Figure 3b - Factor VIII units issued between April 2011 & March 2019 - *Severe Haemophilia A*, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU		IU	% difference since 2017/18	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	2,496,000	1.00	13,630,416	1.00	0				16,126,416	1.00	49	1.00
2012/2013	4,618,000	1.85	12,168,488	0.89	0				16,786,488	1.04	53	1.08
2013/2014	1,988,000	0.80	13,250,855	0.97	0				15,238,855	0.94	57	1.16
2014/2015	754,000	0.30	13,617,750	1.00	0				14,371,750	0.89	57	1.16
2015/2016	753,000	0.30	14,618,000	1.07	0				15,371,000	0.95	60	1.22
2016/2017	834,000	0.33	16,753,000	1.23	0				17,587,000	1.09	66	1.35
2017/2018	1,396,000	0.56	16,462,000	1.21	0		662,000	1.00	18,520,000	1.15	65	1.33
2018/2019	806,500	0.32	17,520,000	1.29	0		1,386,000	2.09	19,712,500	1.22	69	1.41

Table 15c Data for Figure 3c - Factor VIII units issued between April 2011 & March 2019 - *Severe Haemophilia A, Scotland West* postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2012/13	IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	604,000	1.00	10,360,750	1.00	0		0		10,964,750	1.00	54	1.00
2012/2013	525,500	0.87	10,141,500	0.98	105,202	1.00	0		10,772,202	0.98	56	1.04
2013/2014	468,000	0.77	10,878,000	1.05	293,000	2.79	0		11,639,000	1.06	60	1.11
2014/2015	1,169,000	1.94	11,814,500	1.14	314,750	2.99	0		13,298,250	1.21	59	1.09
2015/2016	2,119,500	3.51	13,652,250	1.32	253,500	2.41	0		16,025,250	1.46	61	1.13
2016/2017	2,461,500	4.08	15,598,500	1.51	240,500	2.29	430,750	1.00	18,731,250	1.71	64	1.19
2017/2018	2,063,000	3.42	14,924,500	1.44	0	0.00	1,054,250	2.45	18,041,750	1.65	67	1.24
2018/2019	2,533,500	4.19	14,704,750	1.42	0	0.00	1,723,200	4.00	18,961,450	1.73	68	1.26

Haemophilia B and Factor IX use

Table 16 Factor IX issued, by diagnosis

Coagulation Defect	Patients Treated	FIX (IU)				Total
		Plasma	Recombinant	Enhanced Half-Life	Investigational	
Haemophilia B	66	441,000	3,939,250	2,172,250	-	6,552,500
Haemophilia B Carrier	4	-	60,000	-	-	60,000
Total	70	441,000	3,999,250	2,172,250	-	6,612,500

Table 16 shows the number of people with a Scottish postcode who were issued factor IX concentrate during 2018/19. 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	Total Units
CSL Behring	IDELVION	1,795,750
Grifols	Alphanine	441,000
Novo Nordisk	NovoSeven (mg)	2,915
NovoNordisk	Refixia	108,000
Pfizer	BeneFIX	3,939,250
SOBI/Biogen	ALPROLIX	268,500

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	283,250	70,813
Dundee	5	433,250	86,650
Edinburgh	1	66,000	66,000
Glasgow	14	1,685,000	120,357
	24	2,467,500	102,813

Table 18 reports the number of people with severe haemophilia B treated and the number of units of factor IX issued during 2018/19. This is broken down by the haemophilia centre which issued the treatment.

Note: One person was issued treatment from both Glasgow and Edinburgh Hospitals.

Table 18b Factor IX issued to patients 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	782,500	78,250
Scotland West	14	1,685,000	120,357
	24	2,467,500	102,813

Table 18b reports the number of people with severe haemophilia B treated and the number of units of factor IX issued during 2018/19. This is broken down by region based on the person's postcode as recorded on the NHD.

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	306,070	1	516,000	516,000	1.69
Ayrshire and Arran	369,670	1	156,000	156,000	0.42
Greater Glasgow and Clyde	1,174,980	7	812,750	116,107	0.69
Tayside	416,080	5	433,250	86,650	1.04
Grampian	584,550	4	283,250	70,813	0.48
Highland	321,800	2	121,750	60,875	0.38
Lanarkshire	659,200	3	144,500	48,167	0.22
Borders	115,270	0	-	-	-
Dumfries and Galloway	148,790	0	-	-	-
Fife	371,910	0	-	-	-
Lothian	897,770	0	-	-	-
Shetland	22,990	0	-	-	-
Western Isles	26,830	0	-	-	-
Scotland	5,438,100	23	2,467,500	107,283	0.45

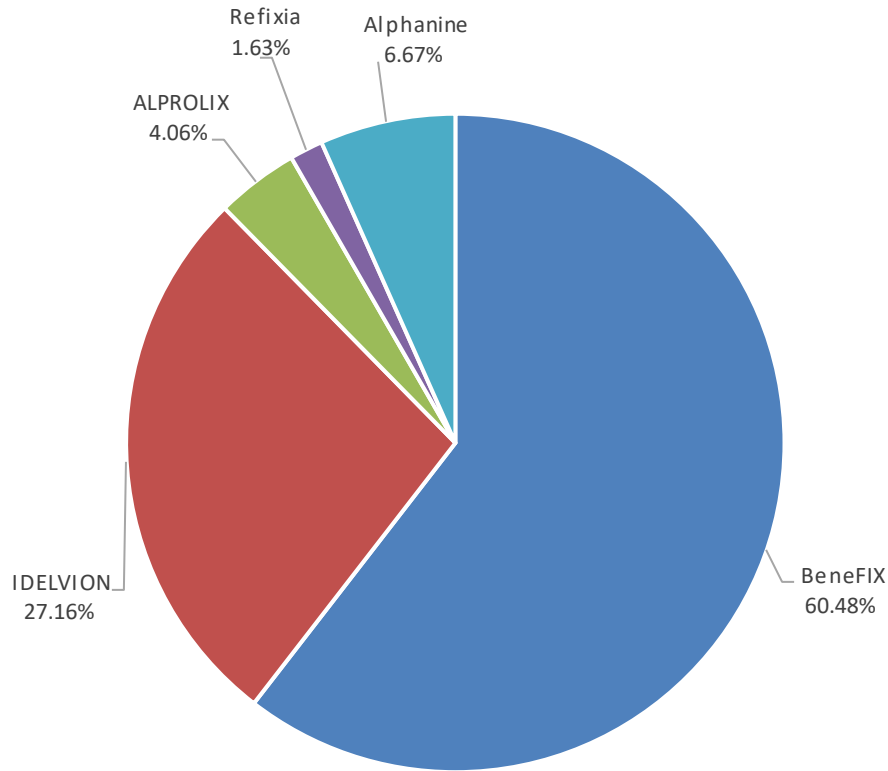
Ranked by mean usage

Mid-2018 population estimates Scotland supplied by National Records of Scotland under the Open Government Licence v3.0 © Crown Copyright 2019

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 patients. People are allocated to a Health Board based on their home postcode.

Figure 4 Market share of factor IX concentrates issued to people with a Scottish postcode between April 2018 & March 2019



Manufacturer	Product	Units (IU)	Patients (n)
Pfizer	BeneFIX	3,999,250	50
CSL Behring	IDELVION	1,795,750	18
SOBI/Biogen	ALPROLIX	268,500	3
NovoNordisk	Refixia	108,000	1
Grifols	Alphanine	441,000	1
	Total	6,612,500	73

Figure 4 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 5a Factor IX units issued between April 2011 & March 2019 - all diagnoses, all severities, all people with a Scottish postcode

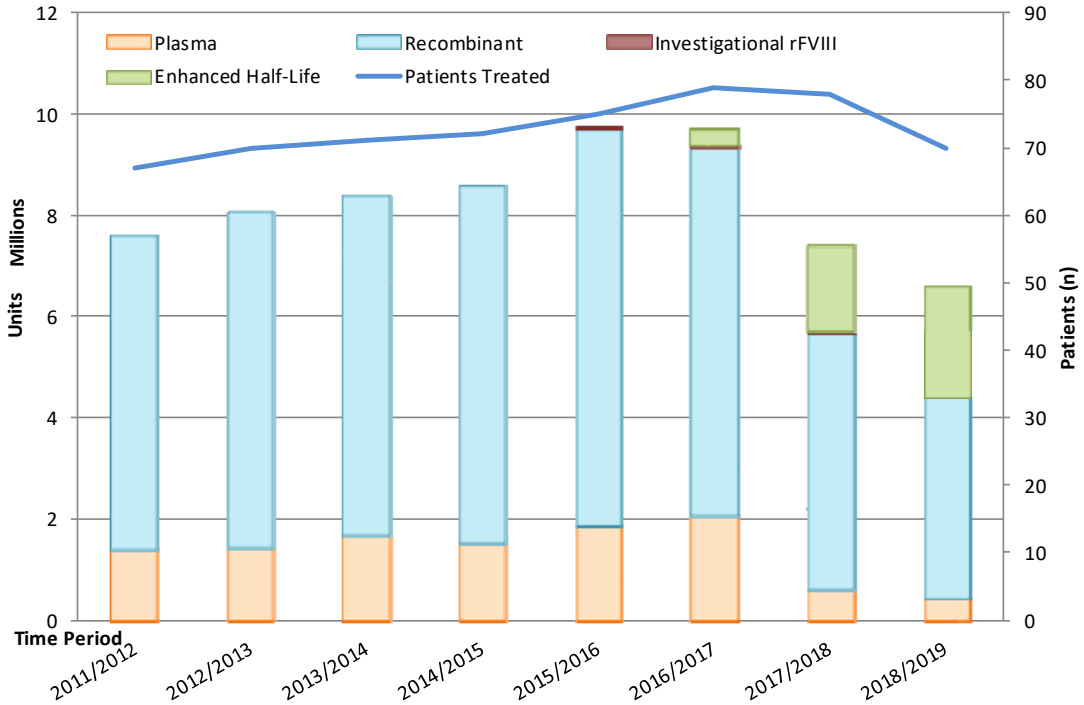


Figure 5b Factor IX units issued between April 2011 & March 2019 - all diagnoses, all severities, people with a Scotland East postcode

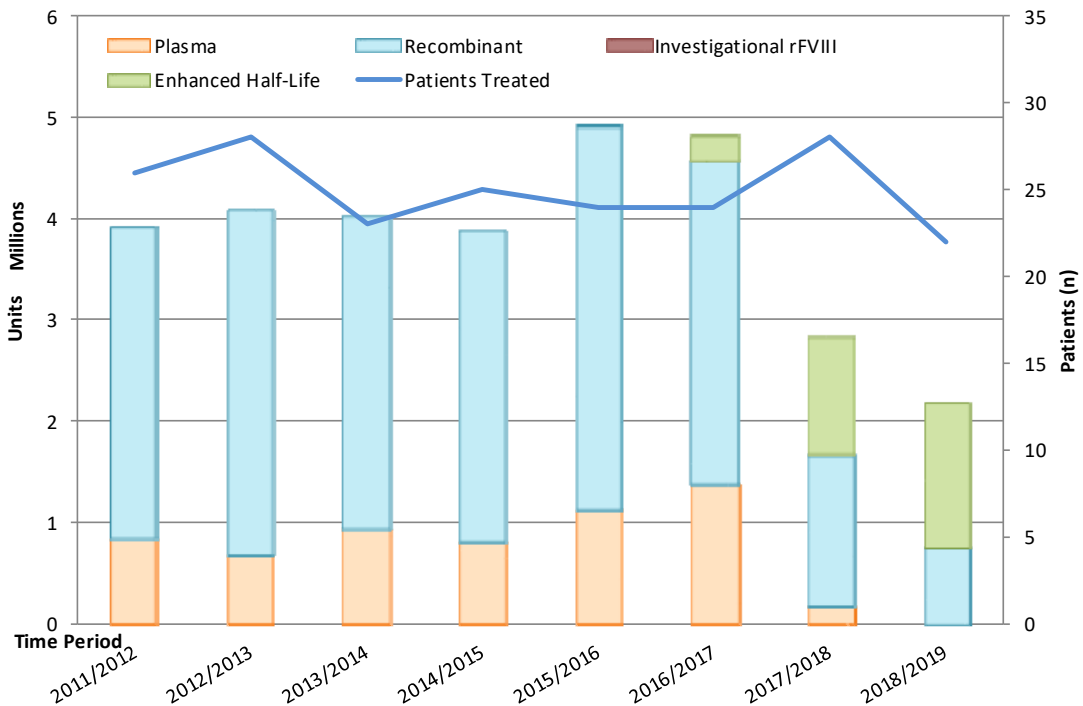
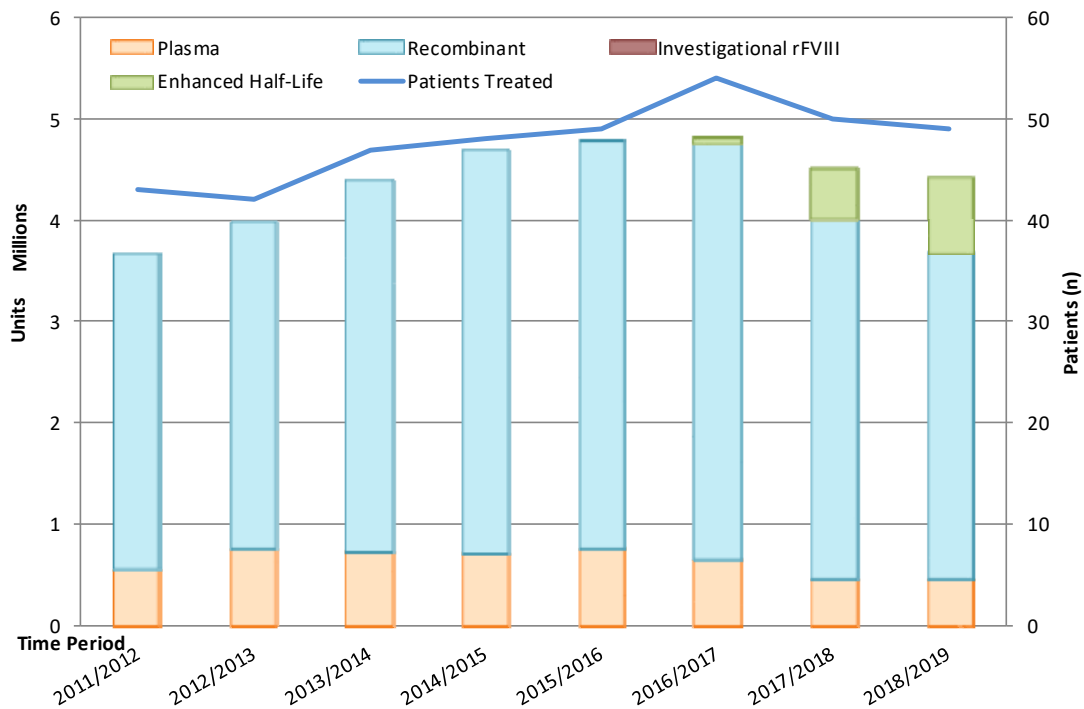


Figure 5c Factor IX units issued between April 2011 & March 2019 - all diagnoses, all severities, people with a Scotland West postcode



Figures 5a to 5c give an historical view of the number of factor IX units issued between 2011/12 and 2018/19 for all diagnoses and all severities. Figure 5a includes all people with a Scottish postcode. Figure 5b includes only people with a Scotland East postcode and Figure 5c 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 5a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 5b and 5c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 5a is not directly comparable to Figures 5b and 5c 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 5 - Factor IX units issued between April 2011 & March 2019 - all diagnoses, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU	% difference since 2015/16	IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	1,386,410	1.00	6,204,779	1.00	0		0		7,591,189	1.00	67	1.00
2012/2013	1,426,250	1.03	6,674,159	1.08	0		0		8,100,409	1.07	70	1.04
2013/2014	1,656,935	1.20	6,767,198	1.09	0		0		8,424,133	1.11	71	1.06
2014/2015	1,501,780	1.08	7,076,500	1.14	0		0		8,578,280	1.13	72	1.07
2015/2016	1,862,370	1.34	7,881,000	1.27	2,500	1.00	0		9,745,870	1.28	75	1.12
2016/2017	2,013,200	1.45	7,345,000	1.18	47,653	19.06	325,500	1.00	9,731,353	1.28	79	1.18
2017/2018	614,305	0.44	5,068,750	0.82	25,588	10.24	1,692,500	5.20	7,401,143	0.97	78	1.16
2018/2019	441,000	0.32	3,999,250	0.64	0	0.00	2,172,250	6.67	6,612,500	0.87	70	1.04

Table 20b Data for figure 5b - Factor IX units issued between April 2011 & March 2019 - all diagnoses, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU		IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	842,910	1.00	3,079,245	1.00	0		0		3,922,155	1.00	26	1.00
2012/2013	667,250	0.79	3,417,909	1.11	0		0		4,085,159	1.04	28	1.08
2013/2014	928,935	1.10	3,092,698	1.00	0		0		4,021,633	1.03	23	0.88
2014/2015	802,780	0.95	3,074,500	1.00	0		0		3,877,280	0.99	25	0.96
2015/2016	1,113,870	1.32	3,813,000	1.24	0		0		4,926,870	1.26	24	0.92
2016/2017	1,366,700	1.62	3,215,500	1.04	0		261,000	1.00	4,843,200	1.23	24	0.92
2017/2018	171,805	0.20	1,487,000	0.48	0		1,181,500	4.53	2,840,305	0.72	28	1.08
2018/2019	0	0.00	748,750	0.24	0		1,436,000	5.50	2,184,750	0.56	22	0.85

Table 20c Data for Figure 5c - Factor IX units issued between April 2011 & March 2019 - all diagnoses, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2011/12	IU	% difference since 2011/12	IU		IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	543,500	1.00	3,125,534	1.00	0		0		3,669,034	1.00	43	1.00
2012/2013	759,000	1.40	3,242,750	1.04	0		0		4,001,750	1.09	42	0.98
2013/2014	728,000	1.34	3,669,500	1.17	0		0		4,397,500	1.20	47	1.09
2014/2015	699,000	1.29	4,002,000	1.28	0		0		4,701,000	1.28	48	1.12
2015/2016	748,500	1.38	4,062,000	1.30	0		0		4,810,500	1.31	49	1.14
2016/2017	646,500	1.19	4,129,500	1.32	0		64,500	1.00	4,840,500	1.32	54	1.26
2017/2018	442,500	0.81	3,581,750	1.15	0		502,750	7.79	4,527,000	1.23	50	1.16
2018/2019	441,000	0.81	3,250,500	1.04	0		736,250	11.41	4,427,750	1.21	49	1.14

Figure 6a Factor IX units issued between April 2011 & March 2019 - *Severe Haemophilia B only*, all people with a Scottish postcode

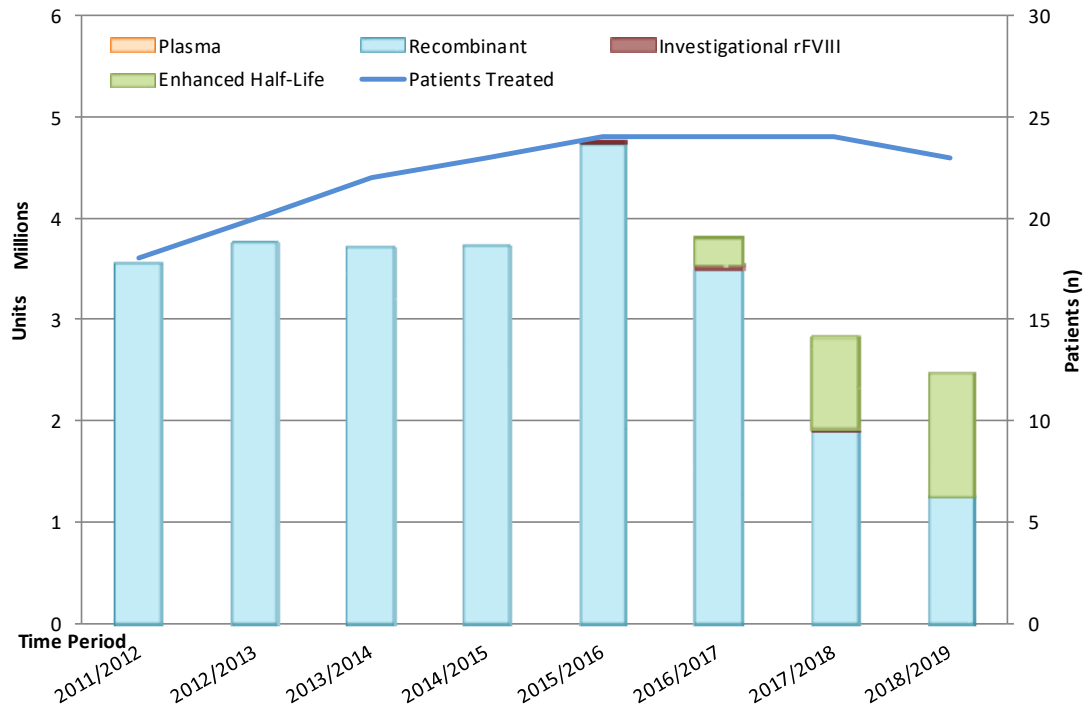


Figure 6b Factor IX units issued between April 2011 & March 2019 - *Severe Haemophilia B only*, people with a Scotland East postcode

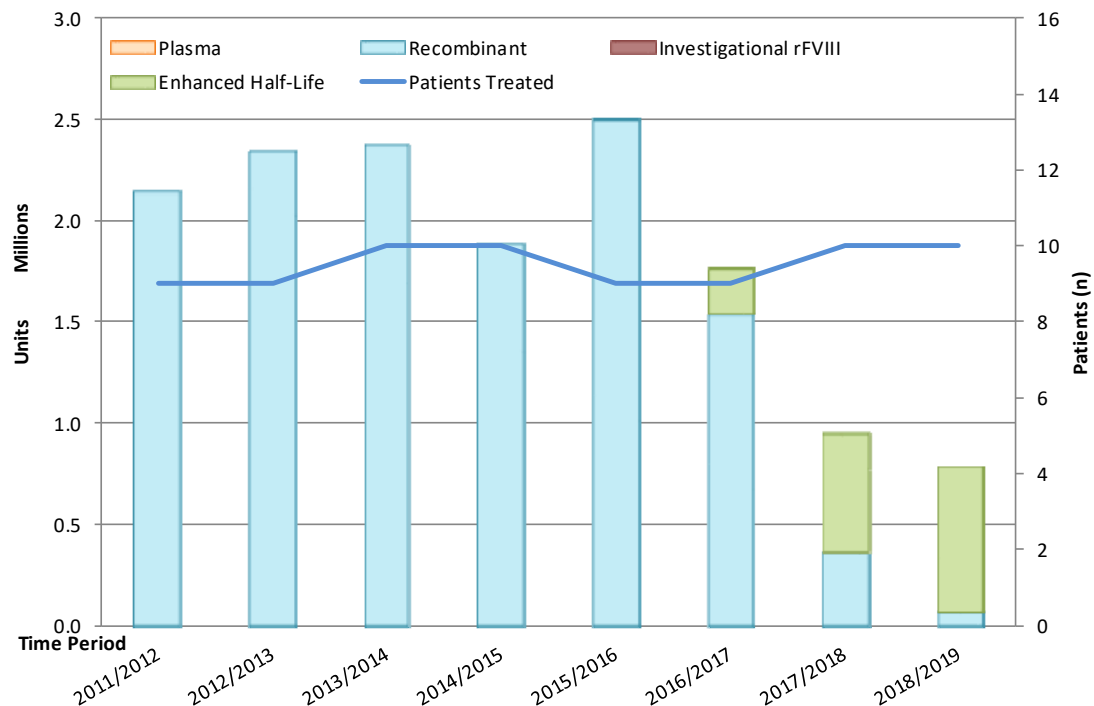
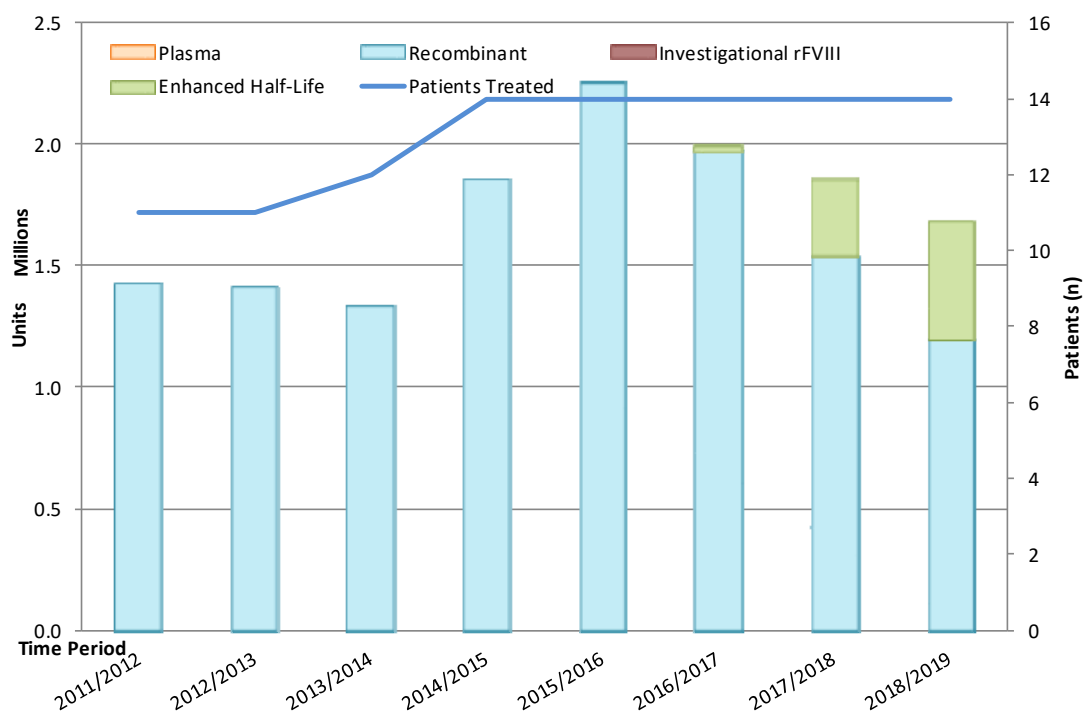


Figure 6c Factor IX units issued between April 2011 & March 2019 - *Severe Haemophilia B only*, people with a Scotland West postcode



Figures 6a to 6c give an historical view of the number of factor IX units issued between 2011/12 and 2018/19 for people with *severe haemophilia B only*. 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 people were issued treatment from both East and West Scotland.

Table 21a Data for Figure 6a - Factor IX units issued between April 2011 & March 2019 - Severe Haemophilia B, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU		IU	% difference since 2011/12	IU	% difference since 2015/16	IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	0		3,577,060	1.00	0		0		3,577,060	1.00	18	1.00
2012/2013	0		3,765,357	1.05	0		0		3,765,357	1.05	20	1.11
2013/2014	0		3,712,792	1.04	0		0		3,712,792	1.04	22	1.22
2014/2015	0		3,744,000	1.05	0		0		3,744,000	1.05	23	1.28
2015/2016	0		4,763,250	1.33	2,500	1.00	0		4,765,750	1.33	24	1.33
2016/2017	0		3,512,000	0.98	47,653	19.06	256,250	1.00	3,815,903	1.07	24	1.33
2017/2018	0		1,899,000	0.53	25,588	10.24	916,500	3.58	2,841,088	0.79	24	1.33
2018/2019	0		1,261,750	0.35	0	0.00	1,205,750	4.71	2,467,500	0.69	23	1.28

Table 21b Data for Figure 6b - Factor IX units issued between April 2011 & March 2019 - Severe Haemophilia B, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU		IU	% difference since 2011/12	IU		IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	0		2,146,060	1.00	0		0		2,146,060	1.00	9	1.00
2012/2013	0		2,350,107	1.10	0		0		2,350,107	1.10	9	1.00
2013/2014	0		2,375,542	1.11	0		0		2,375,542	1.11	10	1.11
2014/2015	0		1,887,000	0.88	0		0		1,887,000	0.88	10	1.11
2015/2016	0		2,502,000	1.17	0		0		2,502,000	1.17	9	1.00
2016/2017	0		1,536,000	0.72	0		230,500	1.00	1,766,500	0.82	9	1.00
2017/2018	0		360,500	0.17	0		585,750	2.54	946,250	0.44	10	1.11
2018/2019	0		66,000	0.03	0		716,500	3.11	782,500	0.36	10	1.11

Table 21c Data for Figure 6c - Factor IX units issued between April 2011 & March 2019 - *Severe Haemophilia B, Scotland West postcodes*

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU		IU	% difference since 2011/12	IU		IU	% difference since 2016/17	IU	% difference since 2011/12	n	% difference since 2011/12
2011/2012	0		1,431,000	1.00	0		0		1,431,000	1.00	11	1.00
2012/2013	0		1,415,250	0.99	0		0		1,415,250	0.99	11	1.00
2013/2014	0		1,337,250	0.93	0		0		1,337,250	0.93	12	1.09
2014/2015	0		1,857,000	1.30	0		0		1,857,000	1.30	14	1.27
2015/2016	0		2,261,250	1.58	0		0		2,261,250	1.58	14	1.27
2016/2017	0		1,976,000	1.38	0		25,750	1.00	2,001,750	1.40	14	1.27
2017/2018	0		1,538,500	1.08	0		322,500	12.52	1,861,000	1.30	14	1.27
2018/2019	0		1,195,750	0.84	0		489,250	19.00	1,685,000	1.18	14	1.27

Von Willebrand Disease, Rarer Bleeding Disorders and Acquired Defects

Tables 21 - 23 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 2018/19, broken down by supplier.

Table 21 Concentrates issued to treat von Willebrand Disease

Manufacturer	Product / Patients (n)	Total Units
CSL Behring	Voncento (n= 51)	712,000
Novo Nordisk	NovoSeven (mg) (n= 1)	232
	Desmopressin (n= 43)	890.0

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

Table 22 Concentrates issued to treat Rarer Bleeding Disorders

Manufacturer	Product / Patients (n)	F.VII Deficiency	F.X Deficiency	F.XI Deficiency	F.XIII Deficiency
BPL	FXI (n= 3)	-	-	4,570	-
CSL Behring	Beriplex (n= 1)	-	2,000	-	-
	Fibrogammin P (n= 3)	-	-	-	49,250
LFB Biomedicaments	Hemoleven (n= 2)	-	-	6,000	-
Novo Nordisk	NovoSeven (mg) (n= 8)	20	-	-	-
	NovoThirteen (n= 1)	-	-	-	35,000

Units in IU unless otherwise stated

Table 23 Concentrates issued to treat Acquired Defects

Manufacturer	Product / Patients (n)	Acquired Haemophilia A	Acquired von Willebrands
CSL Behring	Voncento (n= 3)	-	24,500
Novo Nordisk	NovoSeven (mg) (n= 1)	30	-
Takeda	FEIBA (n= 3)	128,000	-

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

Adverse Events and Deaths

Table 24 Inhibitors by disease severity

Coagulation Defect	Severity (iu/dl) / Subtype	In Register *	Inhibitors		
			New n (%)	Ongoing n (%)	Historical n (%)
Haemophilia A	< 1	146	1 (0.7)	17 (11.6)	34 (23.3)
	1 - 5	70	0 (0.0)	2 (2.9)	6 (8.6)
	> 5	270	0 (0.0)	0 (0.0)	2 (0.7)
	Total	486	1 (0.2)	19 (3.9)	42 (8.6)
Haemophilia B	< 1	25	0 (0.0)	1 (4.0)	1 (4.0)
	1 - 5	43	0 (0.0)	0 (0.0)	0 (0.0)
	> 5	59	0 (0.0)	0 (0.0)	0 (0.0)
	Total	127	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	1,128	0 (0.0)	0 (0.0)	0 (0.0)
	Total	1,158	0 (0.0)	1 (0.1)	1 (0.1)

* Including patients not regularly treated

Table 24 shows the incidence of new inhibitors during 2018/19, 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 2018/19. 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 25 Products issued to people with congenital bleeding disorders reported to have a positive inhibitor during 2018/19

Manufacturer	Product / Patients (n)	Units	Patients
Haemophilia A			
CSL Behring	Helixate Nexgen (n= 1)	34,000	1
Grifols	Fanhdi (n= 2)	2,932,000	2
Novo Nordisk	NovoEight (n= 2)	337,000	2
	NovoSeven (mg) (n= 9)	684	9
Pfizer	ReFacto AF (n= 4)	743,250	4
Roche	Hemlibra (mg) (n= 7)	22,463	7
Takeda	Advate (n= 2)	787,000	2
	FEIBA (n= 6)	3,121,000	6
	Investigational (n= 1)	400	1
Haemophilia B			
Novo Nordisk	NovoSeven (mg) (n= 1)	2,915	1
von Willebrand Disease			
Novo Nordisk	NovoSeven (mg) (n= 1)	232	1
F.XI Deficiency			
LFB Biomedicaments	FXI (n= 1)	1,000	1
	Hemoleven (n= 1)	4,000	1
Co-inherited diagnoses			
Roche	Hemlibra (mg) (n= 1)	5,880	1

Units in IU unless otherwise stated

Table 25 shows the number of people with Scottish postcodes and reported products issued to those with an inhibitor, newly reported or ongoing during 2018/19, broken down by diagnosis and supplier.

Table 26 Adverse Events

Adverse Event	Number of Patients	Number of Events
Allergy Event	0	0
Infection Event	0	0
Inhibitor Event	2	2
Intracranial Haemorrhage	6	7
Malignancy Event	10	11
Neurological Event	0	0
Other Event	0	0
Poor Efficacy Event	0	0
Thrombotic Event	8	10
Total	26	30

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 in patients with a Scottish postcode during 2018/19.

Table 27 Causes of Death

Coagulation Defect	Cause of Death	Severity (factor level iu/dl)			
		< 1	1 - 5	> 5	Total
Haemophilia A	Cerebral haemorrhage	0	1	2	3
	Hepatocellular Carcinoma	1	1	0	2
	Lymphoproliferative Malignancy	1	0	0	1
Acquired Haemophilia A	AIDS	1	0	0	1
	Cerebral haemorrhage	1	0	0	1
	Infection (Bacterial)	1	0	1	2
	Ischaemic Heart Disease	0	0	1	1
	Ruptured Aorta (Peripheral vascular disease)	0	0	1	1
	Unknown	0	0	1	1
von Willebrand disease	Carcinoma				2
	COAD				1
	Infection (Bacterial)				1
Probable von Willebrands disease	Infection (Bacterial)				1
Acquired von Willebrands	ARDS				1
	Unknown				1
F.VII deficiency	Infection (Bacterial)				1
F.XI Deficiency	Accident				1
	Infection (Bacterial)				1
Hypodysfibrinogenemia	Infection (Bacterial)				1
Total		5	2	6	24

Table 28 shows the causes of death reported in people with a Scottish postcode during 2018/19, broken down by diagnosis and disease severity.