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

Bleeding Disorder Statistics for Scotland

April 2017 to March 2018

A report from the UK National Haemophilia Database

April 2019

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 patients newly registered between April 2017 & March 2018, by diagnosis and gender

Coagulation Defect	Male	Female	Total
Haemophilia A	9	1	10
Haemophilia A Carrier		22	22
Acquired Haemophilia A	1	3	4
Haemophilia B	3	0	3
Haemophilia B Carrier		2	2
von Willebrand disease	16	22	38
Probable von Willebrands disease	0	2	2
Acquired von Willebrands	1	0	1
F.VII deficiency	16	18	34
F.X deficiency	2	2	4
F.XI Deficiency	9	15	24
F.XIII Deficiency	0	1	1
Co-inherited diagnoses	0	1	1
Dysfibrinogenemia	4	10	14
Hypofibrinogenemia	2	3	5
Acquired Deficiency (other)	1	0	1
Glanzmanns Thrombasthenia	0	2	2
Platelet defects	3	7	10
Miscellaneous	0	3	3
Unclassified	2	9	11
Total	**69**	**123**	**192**

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 patients with a Scottish postcode.

Table 2 New Registrations of Haemophilia A & B between April 2017 & March 2018, 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	5	0	1	6
	10 : 19	0	0	2	2
	20 : 29	0	0	0	0
	30 : 39	0	0	0	0
	40 : 49	0	0	1	1
	50 : 59	0	0	1	1
	60 : 69	0	0	0	0
	70 +	0	0	0	0
Total		**5**	**0**	**5**	**10**
Haemophilia B	0 : 9	0	1	1	2
	10 : 19	0	0	1	1
	20 : 29	0	0	0	0
	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**	**1**	**2**	**3**

N.B Age calculated at mid-year, 30/09/2017

Table 2 shows the number of new registrations of patients 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 patients in the register as of 31st March 2018 and the number treated between April 2017 & March 2018

Coagulation Defect	In Register			Treated (n)	Treated %
	Males	Females	Total		
Haemophilia A	470	1	471	288	61.15%
Acquired Haemophilia A	18	26	44	3	6.82%
Haemophilia A Carrier		240	240	19	7.92%
Haemophilia A with Liver Transplant	3	0	3	0	0.00%
Haemophilia B	124	0	124	70	56.45%
Haemophilia B Carrier		65	65	8	12.31%
Haemophilia B with Liver Transplant	1	0	1	0	0.00%
von Willebrand disease	396	766	1,162	101	8.69%
Acquired von Willebrands	8	12	20	4	20.00%
Probable von Willebrands disease	16	42	58	8	13.79%
Prothrombin Deficiency	1	2	3	0	0.00%
F.V deficiency	7	13	20	0	0.00%
F.VII deficiency	87	109	196	9	4.59%
F.X deficiency	14	29	43	0	0.00%
F.XI Deficiency	83	117	200	2	1.00%
F.XIII Deficiency	2	2	4	4	100.00%
Combined V+VIII Deficiency	1	2	3	1	33.33%
Co-inherited diagnoses	7	14	21	2	9.52%
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	76	123	199	5	2.51%
Hypofibrinogenemia	8	13	21	0	0.00%
Hypodysfibrinogenemia	7	5	12	0	0.00%
Glanzmanns Thrombasthenia	3	9	12	4	33.33%
Bernard Soulier	3	4	7	0	0.00%
Platelet Defect	50	137	187	4	2.14%
Miscellaneous	8	23	31	2	6.45%
Unclassified	5	40	45	3	6.67%
Totals	**1,399**	**1,799**	**3,198**	**537**	

Table 3 shows the total number of active registrations of patients with a Scottish postcode and the number who were issued treatment during 2017/18.

Table 4 In Register – The total number of patients in the register as of 31st March 2018, by diagnosis and registered Haemophilia Centre

Coagulation Defect	Aberdeen	Dundee	Edinburgh	Glasgow	Inverness	Total
Haemophilia A	69	46	124	240	29	508
Haemophilia A Carrier	28	23	33	153	6	243
Haemophilia A with Liver Transplant	1	1	1	0	0	3
Acquired Haemophilia A	10	2	16	19	0	47
Haemophilia B	8	12	30	74	2	126
Haemophilia B Carrier	1	13	14	40	0	68
Haemophilia B with Liver Transplant	0	1	0	0	0	1
von Willebrand disease	165	189	158	665	40	1217
Acquired von Willebrands	2	3	9	9	0	23
Probable von Willebrands disease	1	34	6	18	0	59
F.V deficiency	3	0	3	14	0	20
F.VII deficiency	35	53	24	87	0	199
F.X deficiency	8	5	6	24	0	43
F.XI Deficiency	17	47	50	84	3	201
F.XIII Deficiency	1	0	1	2	0	4
Combined V+VIII Deficiency	0	0	1	2	0	3
Co-inherited diagnoses	1	6	2	11	0	20
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	2	20	34	145	1	202
Hypofibrinogenemia	4	0	2	14	2	22
Hypodysfibrinogenemia	0	0	11	1	0	12
Glanzmanns Thrombasthenia	1	3	6	3	0	13
Bernard Soulier	1	1	2	3	0	7
Platelet Defect	4	17	91	76	0	188
Miscellaneous	2	9	2	18	0	31
Unclassified	0	1	3	42	0	46
Total	**365**	**486**	**631**	**1747**	**83**	**3,312**

Table 4 shows the number of patients registered at each Haemophilia Centre. Patients are to their registered haemophilia centre and not by their home postcode.

Table 5 In Register – The total number of patients with Haemophilia A & B in the register as of 31st March 2018, 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	51	11	58	**120**
	≥18 years	96	56	199	**351**
	Total	**147**	**67**	**257**	**471**
Haemophilia B	<18 years	7	9	10	**26**
	≥18 years	17	34	47	**98**
	Total	**24**	**43**	**57**	**124**

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

Table 6 In Register – The number of patients with selected rarer bleeding disorders in the register as of 31st March 2018 and the number treated between April 2017 & March 2018, 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	19	0	0	0	20	0
F.VII deficiency	6	2	190	7	0	0	196	9
F.X deficiency	0	0	43	0	0	0	43	0
F.XI Deficiency	11	0	188	2	1	0	200	2
Total	**18**	**2**	**440**	**9**	**1**	**-**	**459**	**11**

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

Table 6 shows the number of patients with selected rarer bleeding disorders and a Scottish postcode known to the NHD during 2017/18. 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 patients with Von Willebrand Disease in the register as of 31st March 2018 and the number treated between April 2017 & March 2018, 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	2	12	9	0	**23**	0	34	68	0	**102**	**125**	**12**
Type 2A	1	0	0	0	**1**	1	5	0	0	**6**	**7**	**2**
Type 2B	0	0	0	0	**0**	0	2	1	0	**3**	**3**	**1**
Type 2M	0	2	0	0	**2**	0	2	2	0	**4**	**6**	**2**
Type 2N	0	0	0	0	**0**	0	0	1	0	**1**	**1**	**0**
Type 2 Unspecified	0	0	1	0	**1**	1	1	5	0	**7**	**8**	**0**
Type 3	1				**1**	6				**6**	**7**	**5**
Type Unreported	6	23	41	0	**70**	10	48	108	0	**166**	**236**	**18**
Low VWF	0	0	1	0	**1**	0	0	2	0	**2**	**3**	**0**
										Sub Total Males	**396**	**40**
Females												
Type 1	0	7	9	0	**16**	2	74	175	0	**251**	**267**	**20**
Type 2A	1	0	0	0	**1**	7	8	1	0	**16**	**17**	**7**
Type 2B	0	0	1	0	**1**	0	5	4	0	**9**	**10**	**1**
Type 2M	1	2	0	0	**3**	6	12	0	0	**18**	**21**	**3**
Type 2N	0	0	0	0	**0**	1	2	0	0	**3**	**3**	**0**
Type 2 Unspecified	0	1	0	0	**1**	0	5	2	0	**7**	**8**	**0**
Type 3	1				**1**	4				**4**	**5**	**1**
Type Unreported	8	22	28	0	**58**	19	86	265	1	**371**	**429**	**29**
Low VWF	0	0	0	0	**0**	0	0	6	0	**6**	**6**	**0**
										Sub Total Females	**766**	**61**
										Grand Total - Males and Females	**1,162**	**101**

Table 7 shows patients 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 Patients with a Scottish postcode, treated between April 2017 & March 2018 and region which issued the treatment, by diagnosis, all severities

Coagulation Defect	Region Issuing Treatment	Patients Treated (n)
Haemophilia A	Cumbria, Northumberland & Tyne and Wear	1
	London	1
	Scotland East	134
	Scotland West	153
	Sub total	**289**
Haemophilia A Carrier	Scotland East	6
	Scotland West	13
	Sub total	**19**
Acquired Haemophilia A	Scotland East	1
	Scotland West	2
	Sub total	**3**
Haemophilia B	London	1
	Scotland East	26
	Scotland West	44
	Sub total	**71**
Haemophilia B Carrier	Scotland West	6
	Scotland East	2
	Sub total	**8**
von Willebrand disease	Birmingham & Black Country	1
	Scotland East	47
	Scotland West	53
	Sub total	**101**
Probable von Willebrand disease	Scotland East	7
	Scotland West	1
	Sub total	**8**
Acquired von Willebrands	Scotland East	4
	Sub total	**4**
F.VII deficiency	Scotland West	3
	Scotland East	6
	Sub total	**9**
F.XI Deficiency	Scotland West	1
	Scotland East	1
	Sub total	**2**
F.XIII Deficiency	Scotland East	2
	Scotland West	2
	Sub total	**4**

Continued overleaf....

Table 8 continued…

Coagulation Defect	Region	Patients Treated (n)
Combined V+VIII Deficiency	Scotland West	1
	Sub total	**1**
Co-inherited diagnoses	Scotland East	1
	Scotland West	1
	Sub total	**2**
Dysfibrinogenemia	Scotland East	1
	Scotland West	4
	Sub total	**5**
Glanzmann's Thrombasthenia	Scotland East	3
	Scotland West	1
	Sub total	**4**
Other platelet defects	Scotland East	4
Miscellaneous	Scotland East	2
Unclassified bleeding disorder	Scotland West	3
	Grand total	**539**

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

Table 8 reports patients with a Scottish postcode by region which issued the treatment. Some patients 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 Patients with a non-Scottish postcode, registered & treated at a Scottish Haemophilia Centre between April 2017 & March 2018, by diagnosis, all severities

Coagulation Defect	Patient's home postcode region	Patients Registered (n)	Patients Treated (n)
Haemophilia A	London	5	3
	North East	1	1
	North West	2	1
	South East	9	5
	South West	3	1
	Wales	0	1
	Sub total	**20**	**12**
Haemophilia A Carrier	East of England	1	0
	North West	1	0
	Sub total	**2**	**0**
Haemophilia B	London	1	2
	Sub total	**1**	**2**
von Willebrand disease	London	3	1
	North East	6	1
	North West	5	0
	Northern Ireland	1	0
	South East	4	1
	Yorkshire and the Humber	3	1
	Sub total	**22**	**4**
Acquired von Willebrands	South East	2	0
	Sub total	**2**	**0**
F.VII deficiency	North East	2	0
	South East	1	0
	Sub total	**3**	**0**
F.XI Deficiency	North West	1	0
	South East	1	0
	Sub total	**2**	**0**
Dysfibrinogenemia	East Midlands	1	0
Glanzmanns Thrombasthenia	North West	1	1
	Grand total	**54**	**19**

The patients reported in Table 9 were registered at or issued treatment from a Scottish Haemophilia Centre during 2017/18, 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)				
		Plasma	Recombinant	Enhanced Half-Life	Investigational	Total
Haemophilia A	267	3,459,000	42,596,750	3,089,750	-	49,145,500
Haemophilia A Carrier	12	-	66,500	-	-	66,500
Acquired Haemophilia A	1	-	12,000	-	-	12,000
von Willebrand disease	58	741,500	-	-	-	741,500
Acquired von Willebrands	4	18,000	-	-	-	18,000
Combined V+VIII Deficiency	1	-	4,000	-	-	4,000
Co-inherited diagnoses	1	1,500	-	-	-	1,500
Total	**344**	**4,220,000**	**42,679,250**	**3,089,750**	**-**	**49,989,000**

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

Table 10 shows the number of patients with a Scottish postcode who were issued factor VIII concentrate during 2017/18. 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	1,460,250
CSL Behring	Helixate Nexgen	1,130,500
Grifols	Fanhdi	3,459,000
Novo Nordisk	NovoEight	217,750
	NovoSeven (mg)	927
Octapharma	Nuwiq	40,000
Pfizer	ReFacto AF	24,239,000
Shire	Advate	15,509,250
	FEIBA	5,253,500
SOBI/Biogen	Elocta	3,089,750

Units in IU unless otherwise stated

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

Table 12a Factor VIII issued to patients 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	20	5,598,000	279,900
Dundee	16	3,946,000	246,625
Edinburgh	29	9,423,500	324,948
Glasgow	69	18,419,750	266,953
Inverness	2	242,500	121,250
Royal Free	1	17,500	17,500
	137	**37,647,250**	**274,797**

Table 12a reports the number of patients with severe haemophilia A treated and the number of units of factor VIII issued during 2017/18. This is broken down by the haemophilia centre which issued the treatment.

Table 12b Factor VIII issued by region for patients 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	67	19,210,000	286,716
Scotland West	69	18,419,750	266,953
London	1	17,500	17,500
	137	**37,647,250**	**274,797**

Table 12b reports the number of patients with severe haemophilia A treated and the number of units of factor VIII issued during 2017/18. This is broken down by region based on the patient's postcode as recorded on the NHD.

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

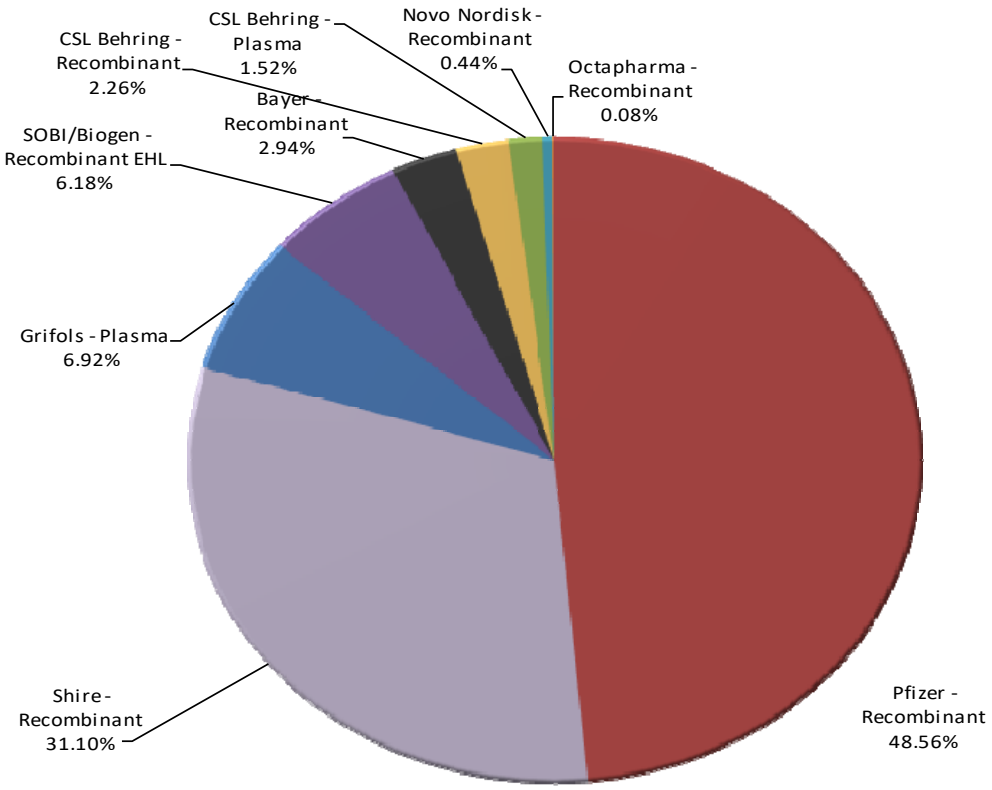
Health Board	General Population	Severe Haemophilia A			
		Patients treated (n)	Total FVIII Units (IU)	Mean Usage	FVIII Units Per Capita
Borders	115,020	2	1,927,000	963,500	16.75
Dumfries and Galloway	149,200	3	1,581,000	527,000	10.60
Forth Valley	305,580	6	2,365,250	394,208	7.74
Lothian	889,450	22	6,446,000	293,000	7.25
Grampian	586,380	19	5,556,250	292,434	9.48
Highland	321,990	5	1,317,750	263,550	4.09
Tayside	416,090	11	2,839,000	258,091	6.82
Lanarkshire	658,130	7	1,761,750	251,679	2.68
Ayrshire and Arran	370,410	11	2,748,500	249,864	7.42
Greater Glasgow and Clyde	1,169,110	38	8,882,500	233,750	7.60
Shetland	23,080	3	560,750	186,917	24.30
Fife	371,410	8	1,412,000	176,500	3.80
Western Isles	26,950	2	249,500	124,750	9.26
Scotland	**5,424,800**	**137**	**37,647,250**	**274,797**	**6.94**

Ranked by mean usage

Table 13 reports the number of patients 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. Patients are allocated to a Health Board based on their home postcode.

Figure 1 Market share of factor VIII concentrates issued between April 2017 & March 2018



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

Manuacturer-Product Type	Patients (n)	FVIII Units (IU)
Pfizer - Recombinant	150	24,276,000
Shire - Recombinant	94	15,547,750
Grifols - Plasma	3	3,459,000
SOBI/Biogen - Recombinant EHL	20	3,089,750
Bayer - Recombinant	10	1,467,250
CSL Behring - Recombinant	17	1,130,500
CSL Behring - Plasma	64	761,000
Novo Nordisk - Recombinant	11	217,750
Octapharma - Recombinant	1	40,000
Total	**370**	**49,989,000**

Figure 1 shows the market breakdown of factor VIII concentrates issued for all diagnoses, including patients with inhibitors. Also included is a table showing the number of patients issued with these products and the number of units issued.

NOTE: The number of patients 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 2010 & March 2018 - all diagnoses, all severities, all patients with a Scottish postcode

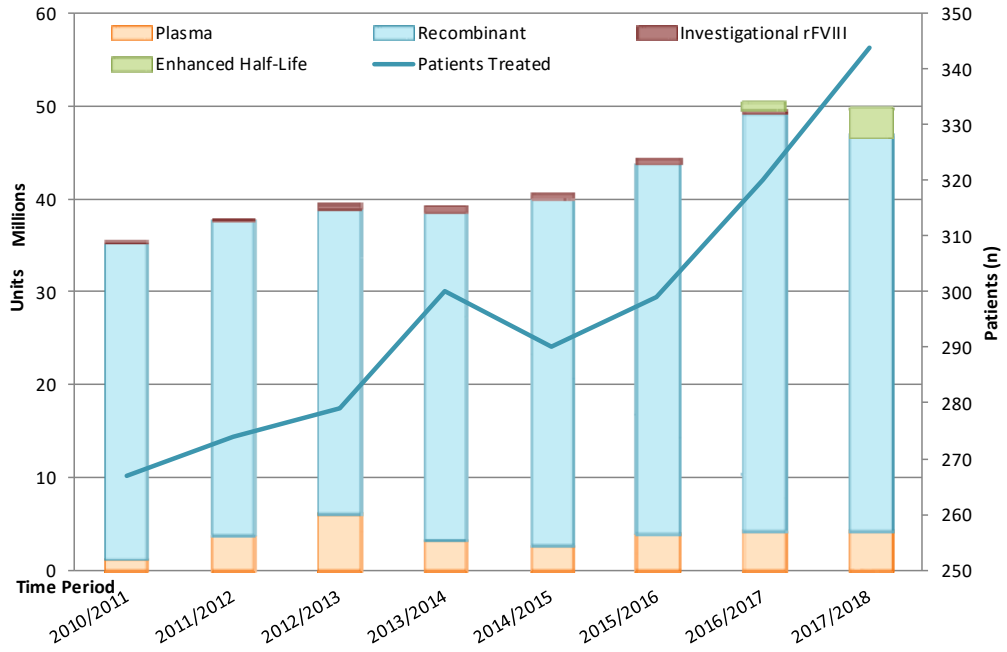


Figure 2b Factor VIII units issued between April 2010 & March 2018 - all diagnoses, all severities, patients with a Scotland East postcode

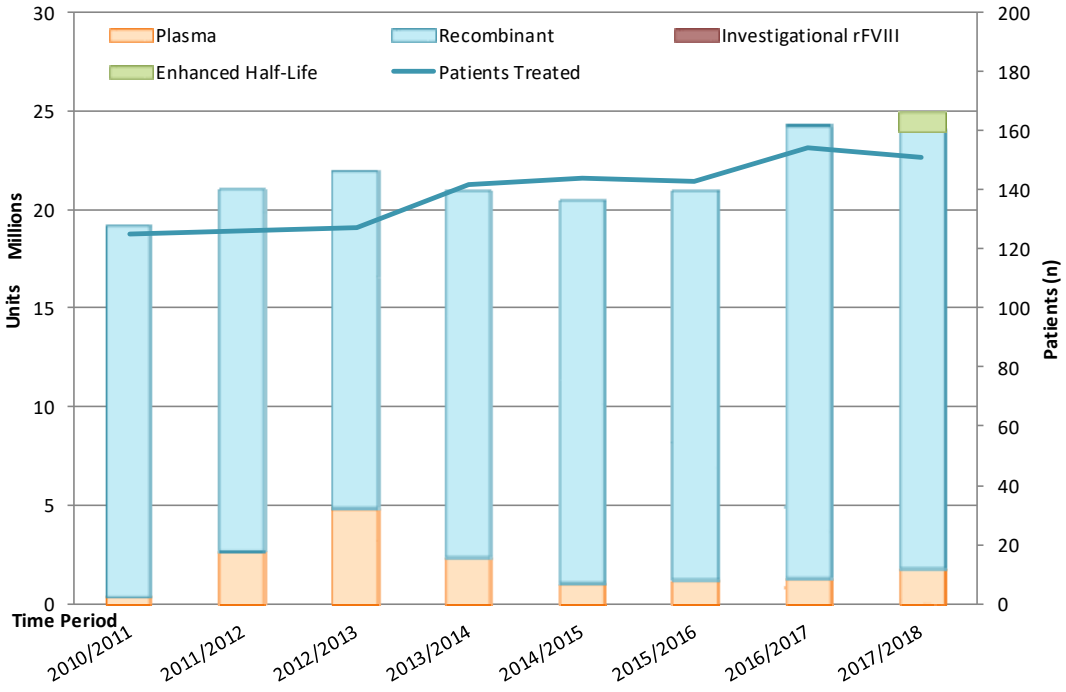
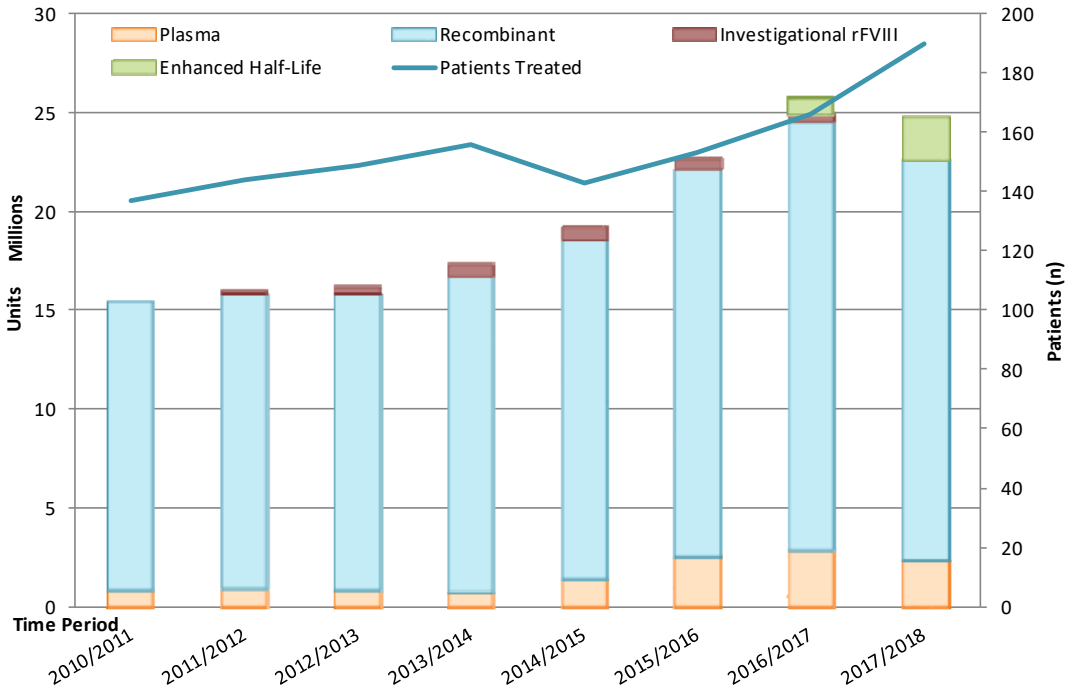


Figure 2c Factor VIII units issued between April 2010 & March 2018 – all diagnoses, all severities, patients with a Scotland West postcode



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 2010/11 and 2017/18 for all diagnoses and all severities. Figure 2a includes all patients with a Scottish postcode. Figures 2b includes only patients with a Scotland East postcode and Figure 2c those with a Scotland West postcode. The number of patients 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 patients 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 patients 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 14a Data for Figure 2a - Factor VIII units issued between April 2010 & March 2018 – all diagnoses, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU	% difference since 2010/11	IU	% difference since 2016/17	IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	1,151,000	1.00	34,452,030	1.00	144,120	1.00	0		35,747,150	1.00	267	1.00
2011/2012	3,614,450	3.14	34,264,069	0.99	139,404	0.97	0		38,017,923	1.06	274	1.03
2012/2013	5,877,400	5.11	33,274,975	0.97	472,202	3.28	0		39,624,577	1.11	279	1.04
2013/2014	3,168,100	2.75	35,710,395	1.04	630,000	4.37	0		39,508,495	1.11	300	1.12
2014/2015	2,602,000	2.26	37,489,500	1.09	674,750	4.68	0		40,766,250	1.14	290	1.09
2015/2016	3,763,550	3.27	40,120,250	1.16	613,500	4.26	0		44,497,300	1.24	299	1.12
2016/2017	4,179,500	3.63	45,244,750	1.31	330,500	2.29	849,750	1.00	50,604,500	1.42	320	1.20
2017/2018	4,220,000	3.67	42,679,250	1.24	0	0.00	3,089,750	3.64	49,989,000	1.40	344	1.29

Table 14b Data for Figure 2b - Factor VIII units issued between April 2010 & March 2018 – all diagnoses, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU		IU		IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	348,500	1.00	18,806,203	1.00	0		0		19,154,703	1.00	125	1.00
2011/2012	2,668,000	7.66	18,481,568	0.98	0		0		21,149,568	1.10	126	1.01
2012/2013	4,846,900	13.91	17,183,225	0.91	0		0		22,030,125	1.15	127	1.02
2013/2014	2,301,100	6.60	18,725,645	1.00	0		0		21,026,745	1.10	142	1.14
2014/2015	1,058,000	3.04	19,394,750	1.03	0		0		20,452,750	1.07	144	1.15
2015/2016	1,170,500	3.36	19,859,500	1.06	0		0		21,030,000	1.10	143	1.14
2016/2017	1,258,000	3.61	23,134,500	1.23	0		0		24,392,500	1.27	154	1.23
2017/2018	1,785,500	5.12	22,300,250	1.19	0		919,000		25,004,750	1.31	151	1.21

Table 14c Data for Figure 2c - Factor VIII units issued between April 2010 & March 2018 – all diagnoses, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU	% difference since 2011/12	IU	% difference since 2016/17	IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	750,500	1.00	14,654,827	1.00	0		0		15,405,327	1.00	137	1.00
2011/2012	856,450	1.14	15,000,501	1.02	137,500	1.00	0		15,994,451	1.04	144	1.05
2012/2013	782,500	1.04	15,032,750	1.03	472,202	3.43	0		16,287,452	1.06	149	1.09
2013/2014	711,000	0.95	16,042,250	1.09	630,000	4.58	0		17,383,250	1.13	156	1.14
2014/2015	1,383,000	1.84	17,221,750	1.18	674,750	4.91	0		19,279,500	1.25	143	1.04
2015/2016	2,469,550	3.29	19,669,750	1.34	613,500	4.46	0		22,752,800	1.48	153	1.12
2016/2017	2,783,500	3.71	21,851,250	1.49	330,500	2.40	849,750	1.00	25,815,000	1.68	166	1.21
2017/2018	2,318,500	3.09	20,357,500	1.39	0	0.00	2,170,750	2.55	24,846,750	1.61	190	1.39

Figure 3a Factor VIII units issued between April 2010 & March 2018 - *Severe Haemophilia A only*, all patients with a Scottish postcode

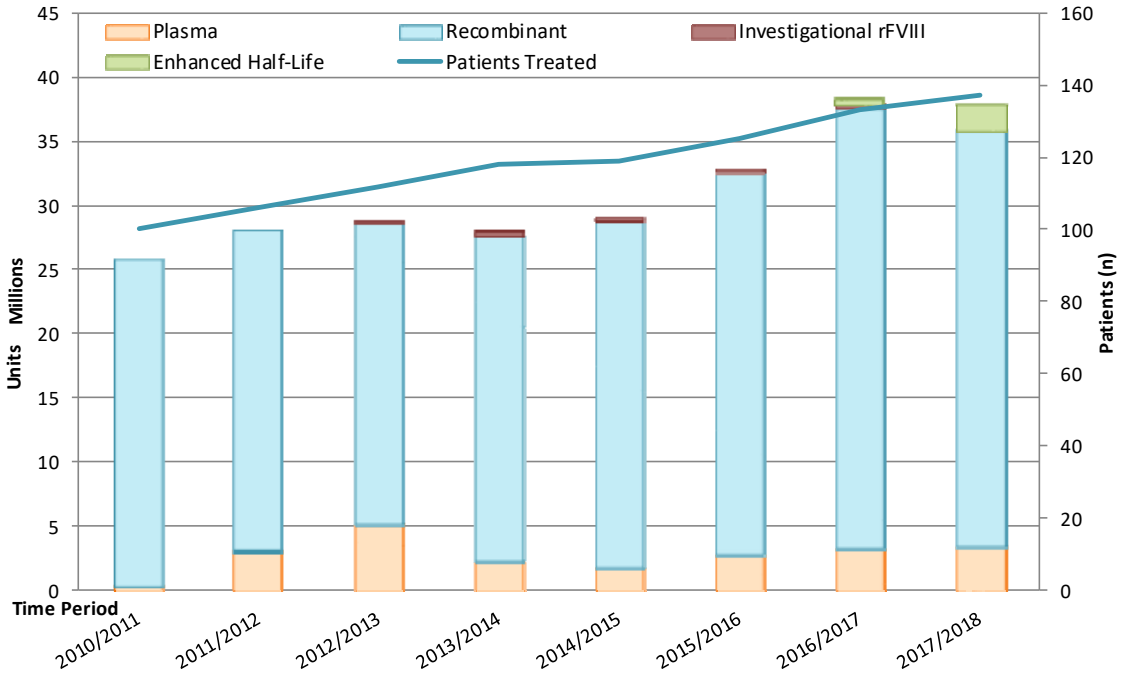


Figure 3b Factor VIII units issued between April 2010 & March 2018 - *Severe Haemophilia A only*, patients with a Scotland East postcode

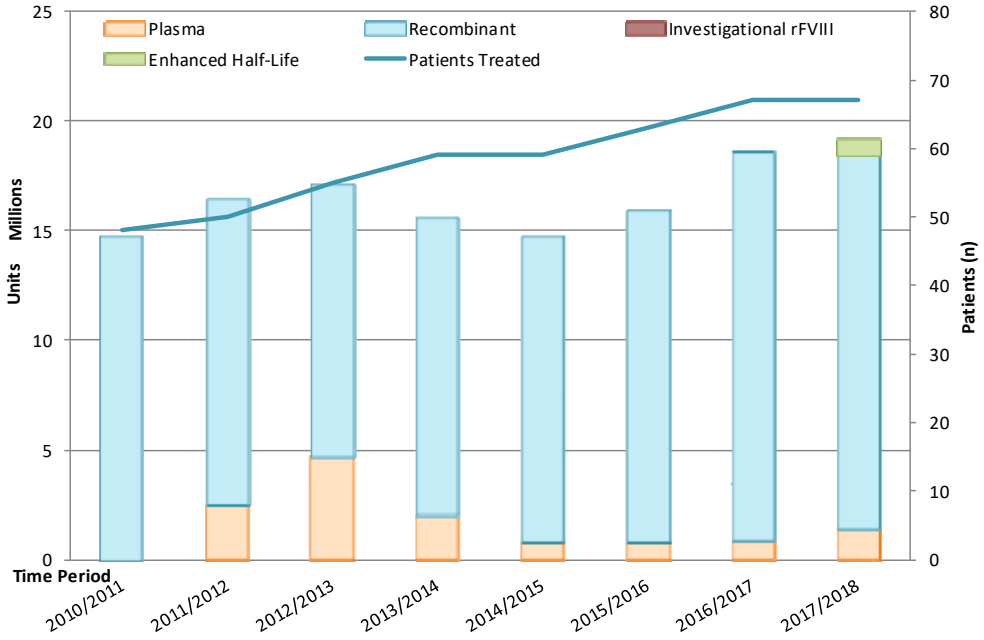
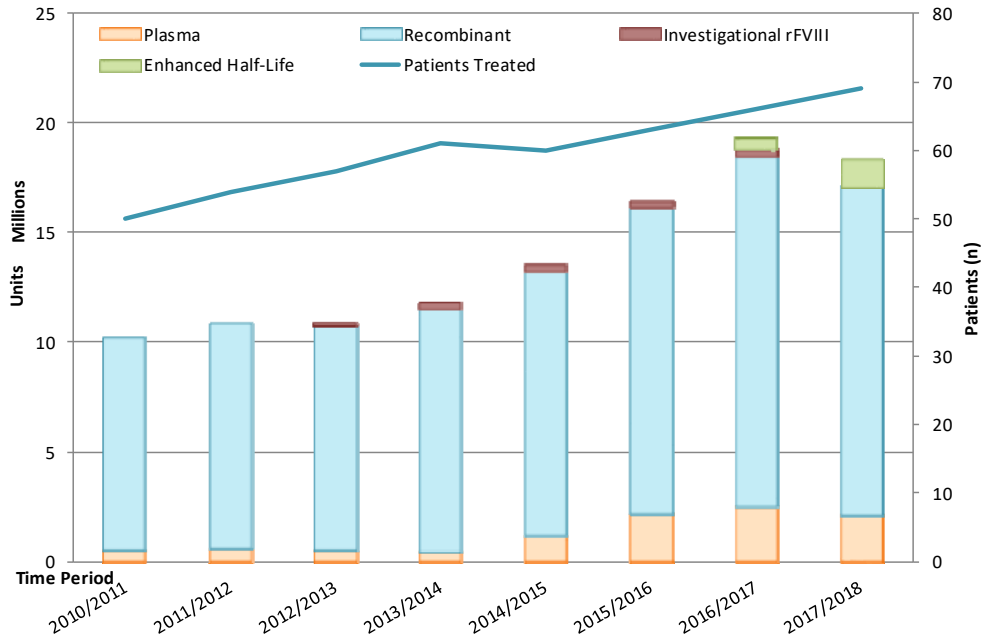


Figure 3c Factor VIII units issued between April 2010 & March 2018 – *Severe Haemophilia A only*, patients with a Scotland West postcode



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 2010/11 and 2017/18 for patients with *severe haemophilia A only*. Figure 3a includes all patients with a Scottish postcode. Figures 3b includes only patients with a Scotland East postcode and Figure 3c those with a Scotland West postcode. The number of patients 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 patients 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 patients 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 15a Data for Figure 3a - Factor VIII units issued between April 2010 & March 2018 - *Severe Haemophilia A*, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since2010/11	IU	% difference since2010/11	IU	% difference since2012/13	IU	% difference since 2016/17	IU	% difference since2010/11	n	% difference since2010/11
2010/2011	512,000	1.00	25,305,201	1.00	0		0		25,817,201	1.00	100	1.00
2011/2012	3,100,000	6.05	24,952,416	0.99	0		0		28,052,416	1.09	106	1.06
2012/2013	5,143,500	10.05	23,589,526	0.93	105,202	1.00	0		28,838,228	1.12	112	1.12
2013/2014	2,456,000	4.80	25,291,355	1.00	293,000	2.79	0		28,040,355	1.09	118	1.18
2014/2015	1,923,000	3.76	26,833,750	1.06	314,750	2.99	0		29,071,500	1.13	119	1.19
2015/2016	2,872,500	5.61	29,724,750	1.17	253,500	2.41	0		32,850,750	1.27	125	1.25
2016/2017	3,295,500	6.44	34,149,750	1.35	240,500	2.29	536,750	1.00	38,222,500	1.48	133	1.33
2017/2018	3,459,000	6.76	32,255,500	1.27	0	0.00	1,932,750	3.60	37,647,250	1.46	137	1.37

Table 15b Data for Figure 3b - Factor VIII units issued between April 2010 & March 2018 - *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 2010/11	IU		IU		IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	0		14,763,304	1.00	0		0		14,763,304	1.00	48	1.00
2011/2012	2,496,000	1.00	13,994,416	0.95	0		0		16,490,416	1.12	50	1.04
2012/2013	4,618,000	1.85	12,519,526	0.85	0		0		17,137,526	1.16	55	1.15
2013/2014	1,988,000	0.80	13,636,855	0.92	0		0		15,624,855	1.06	59	1.23
2014/2015	754,000	0.30	13,984,500	0.95	0		0		14,738,500	1.00	59	1.23
2015/2016	753,000	0.30	15,242,500	1.03	0		0		15,995,500	1.08	63	1.31
2016/2017	834,000	0.33	17,834,000	1.21	0		0		18,668,000	1.26	67	1.40
2017/2018	1,396,000	0.56	17,152,000	1.16	0		662,000		19,210,000	1.30	67	1.40

Table 15c Data for Figure 3c - Factor VIII units issued between April 2010 & March 2018 - *Severe Haemophilia A*, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU	% difference since 2012/13	IU	% difference since 2016/17	IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	512,000	1.00	9,689,897	1.00	0		0		10,201,897	1.00	50	1.00
2011/2012	604,000	1.18	10,286,000	1.06	0		0		10,890,000	1.07	54	1.08
2012/2013	525,500	1.03	10,273,000	1.06	105,202	1.00	0		10,903,702	1.07	57	1.14
2013/2014	468,000	0.91	11,041,000	1.14	293,000	2.79	0		11,802,000	1.16	61	1.22
2014/2015	1,169,000	2.28	12,122,250	1.25	314,750	2.99	0		13,606,000	1.33	60	1.20
2015/2016	2,119,500	4.14	14,040,250	1.45	253,500	2.41	0		16,413,250	1.61	63	1.26
2016/2017	2,461,500	4.81	16,104,750	1.66	240,500	2.29	536,750	1.00	19,343,500	1.90	66	1.32
2017/2018	2,063,000	4.03	15,086,000	1.56	0	0.00	1,270,750	2.37	18,419,750	1.81	69	1.38

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	70	614,305	5,005,750	1,692,500	25,588	7,338,143
Haemophilia B Carrier	8	-	63,000	-	-	63,000
Total	**78**	**614,305**	**5,068,750**	**1,692,500**	**25,588**	**7,401,143**

Table 16 shows the number of patients with a Scottish postcode who were issued factor IX concentrate during 2017/18. 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
BPL	Replenine	171,805
CSL Behring	IDELVION	1,460,750
Grifols	Alphanine	442,500
Pfizer	BeneFIX	5,005,750
SOBI/Biogen	ALPROLIX	231,750
	Investigational	25,588

Units in IU unless otherwise stated

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

Table 18a Factor IX issued to patients 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	351,000	87,750
Dundee	5	591,250	118,250
Edinburgh	1	4,000	4,000
Glasgow	14	1,861,000	132,929
Great Ormond Street	1	33,838	33,838
	25	**2,841,088**	**113,644**

Table 18 reports the number of patients with severe haemophilia B treated and the number of units of factor IX issued during 2017/18. This is broken down by the haemophilia centre which issued the treatment.

Note: One patient 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	946,250	94,625
Scotland West	14	1,861,000	132,929
London	1	33,838	33,838
	25	**2,841,088**	**113,644**

Table 18b reports the number of patients with severe haemophilia B treated and the number of units of factor IX issued during 2017/18. This is broken down by region based on the patient's postcode as recorded on the NHD.

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

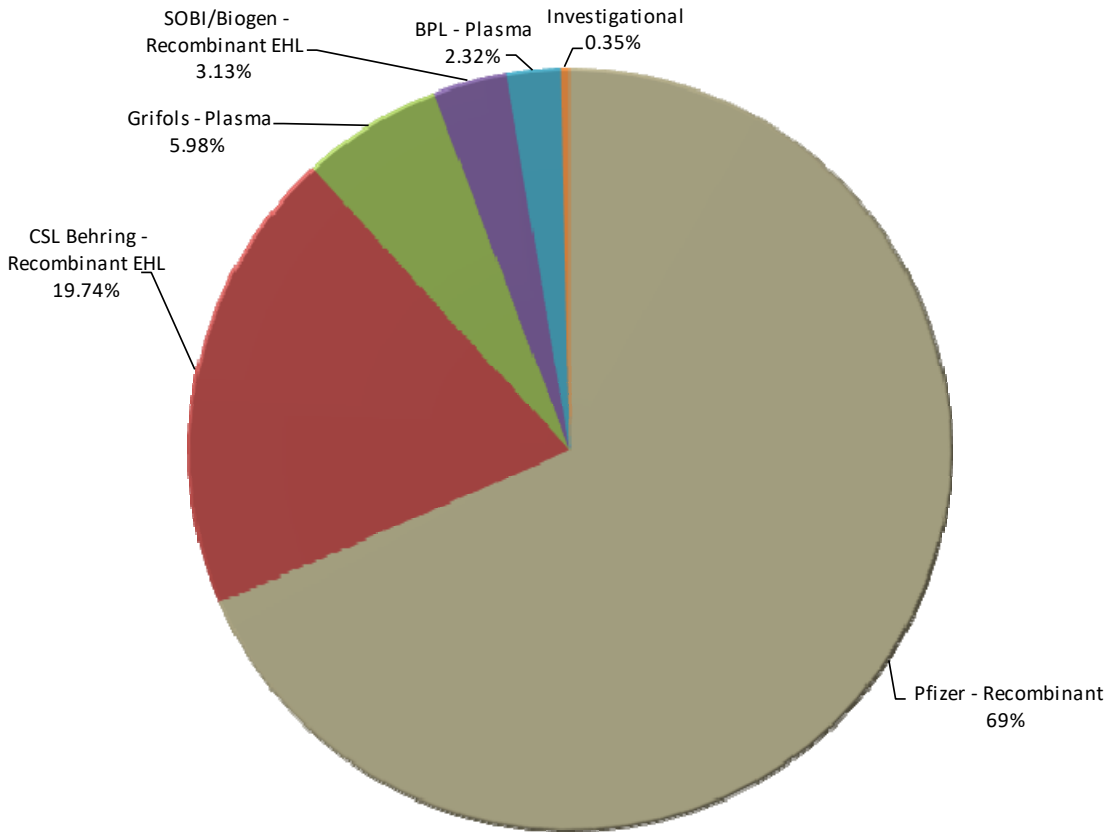
Health Board	General Population	Severe Haemophilia B			
		Patients treated (n)	Total FIX Units (IU)	Mean Usage	FIX Units Per Capita
Forth Valley	305,580	1	400,000	400,000	1.31
Ayrshire and Arran	370,410	1	198,000	198,000	0.53
Greater Glasgow and Clyde	1,169,110	8	1,055,338	131,917	0.90
Tayside	416,090	5	591,250	118,250	1.42
Grampian	586,380	4	351,000	87,750	0.60
Highland	321,990	2	151,000	75,500	0.47
Lanarkshire	658,130	3	94,500	31,500	0.14
Borders	115,020	0	-	-	-
Dumfries and Galloway	149,200	0	-	-	-
Fife	371,410	0	-	-	-
Lothian	889,450	0	-	-	-
Shetland	23,080	0	-	-	-
Western Isles	26,950	0	-	-	-
Scotland	**5,424,800**	**24**	**2,841,088**	**118,379**	**0.52**

Ranked by mean usage

Table 19 reports the number of patients 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 patient. Usage per capita of population is also reported.

This table does not contain duplicate patients. Patients are allocated to a Health Board based on their home postcode.

Figure 4 Market share of factor IX concentrates issued to patients with a Scottish postcode between April 2017 & March 2018



Manuacturer-Product Type	Patients (n)	FIX Units (IU)
Pfizer - Recombinant	63	5,068,750
CSL Behring - Recombinant EHL	16	1,460,750
Grifols - Plasma	1	442,500
SOBI/Biogen - Recombinant EHL	3	231,750
BPL - Plasma	1	171,805
Investigational	1	25,588
Total	**85**	**7,401,143**

Figure 4 shows the market breakdown of factor IX concentrates issued for all diagnoses, including patients with inhibitors. Also included is a table showing the number of patients issued with these products and the number of units issued.

NOTE: The number of patients in this table cannot be compared with table 16 as this table includes patients 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 2010 & March 2018 – all diagnoses, all severities, all patients with a Scottish postcode

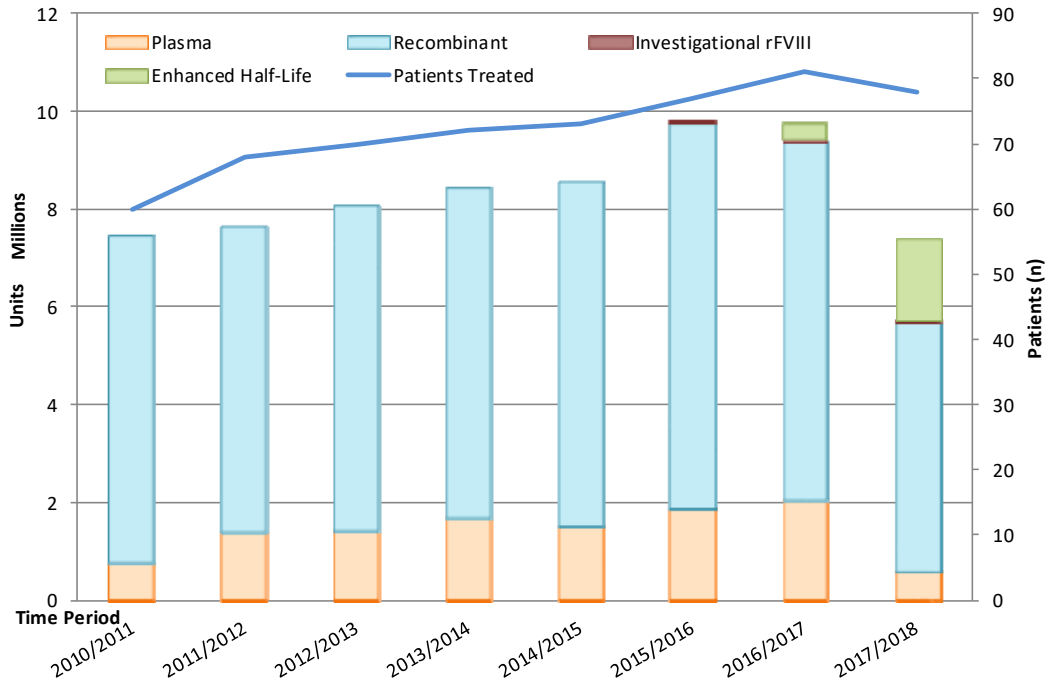


Figure 5b Factor IX units issued between April 2010 & March 2018 – all diagnoses, all severities, patients with a Scotland East postcode

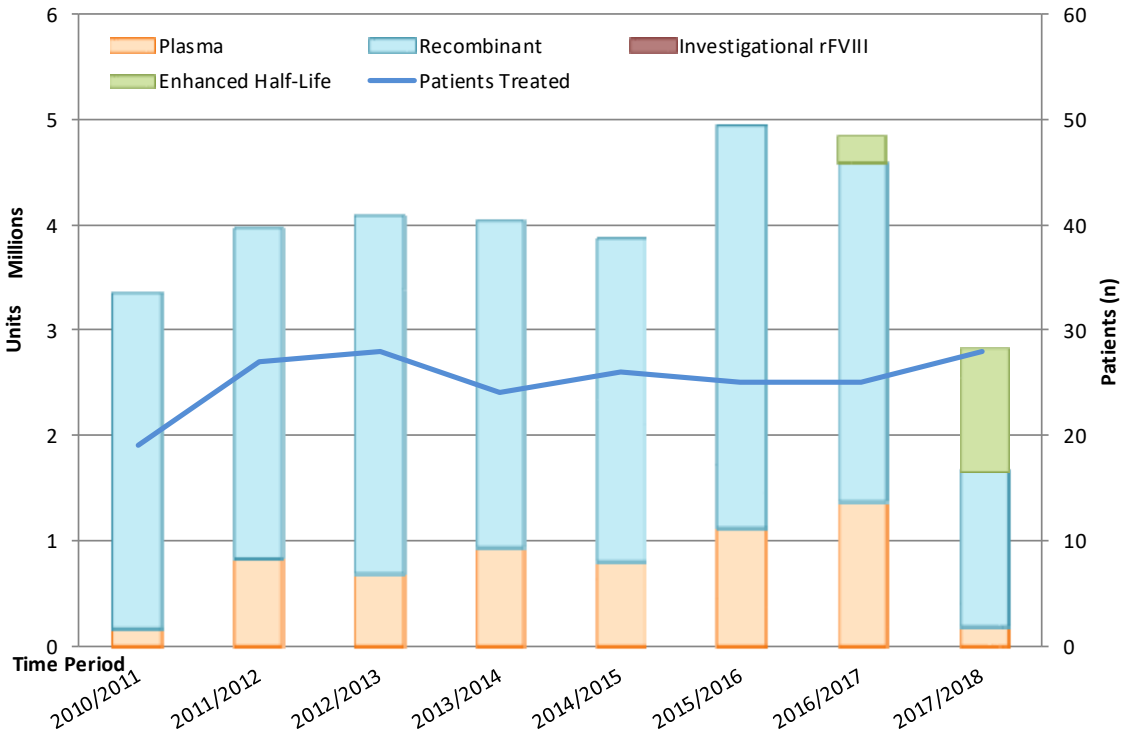
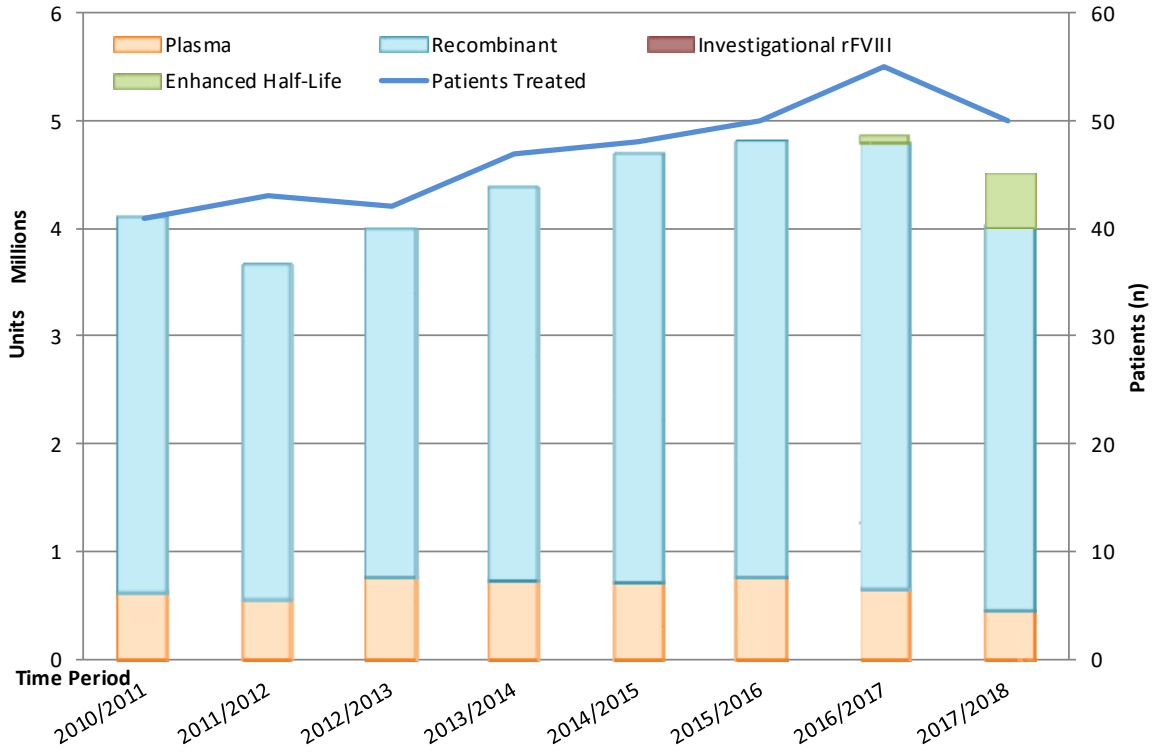


Figure 5c Factor IX units issued between April 2010 & March 2018 – all diagnoses, all severities, patients with a Scotland West postcode



Figures 5a to 5c give an historical view of the number of factor IX units issued between 2010/11 and 2017/18 for all diagnoses and all severities. Figure 5a includes all patients with a Scottish postcode. Figures 5b includes only patients with a Scotland East postcode and Figure 5c those with a Scotland West postcode. The number of patients 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 patients 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 patients 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 2010 & March 2018 – all diagnoses, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU	% difference since 2015/16	IU	% difference since 2016/17	IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	766,570	1.00	6,708,083	1.00	0		0		7,474,653	1.00	60	1.00
2011/2012	1,386,410	1.81	6,267,181	0.93	0		0		7,653,591	1.02	68	1.13
2012/2013	1,426,250	1.86	6,674,159	0.99	0		0		8,100,409	1.08	70	1.17
2013/2014	1,656,935	2.16	6,792,240	1.01	0		0		8,449,175	1.13	72	1.20
2014/2015	1,501,780	1.96	7,079,500	1.06	0		0		8,581,280	1.15	73	1.22
2015/2016	1,862,370	2.43	7,904,000	1.18	2,500	1.00	0		9,768,870	1.31	77	1.28
2016/2017	2,013,200	2.63	7,381,000	1.10	47,653	19.06	325,500	1.00	9,767,353	1.31	81	1.35
2017/2018	614,305	0.80	5,068,750	0.76	25,588	10.24	1,692,500	5.20	7,401,143	0.99	78	1.30

Table 20b Data for figure 5b - Factor IX units issued between April 2010 & March 2018 – all diagnoses, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU		IU	% difference since 2016/17	IU	% difference since 2010/11	n	% differ since 201
2010/2011	162,670	1.00	3,195,124	1.00	0		0		3,357,794	1.00	19	
2011/2012	842,910	5.18	3,141,647	0.98	0		0		3,984,557	1.19	27	
2012/2013	667,250	4.10	3,417,909	1.07	0		0		4,085,159	1.22	28	
2013/2014	928,935	5.71	3,117,740	0.98	0		0		4,046,675	1.21	24	
2014/2015	802,780	4.94	3,077,500	0.96	0		0		3,880,280	1.16	26	
2015/2016	1,113,870	6.85	3,831,000	1.20	0		0		4,944,870	1.47	25	
2016/2017	1,366,700	8.40	3,221,500	1.01	0		261,000	1.00	4,849,200	1.44	25	
2017/2018	171,805	1.06	1,487,000	0.47	0		1,181,500	4.53	2,840,305	0.85	28	

Table 20c Data for Figure 5c - Factor IX units issued between April 2010 & March 2018 – all diagnoses, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU	% difference since 2010/11	IU	% difference since 2010/11	IU		IU	% difference since 2016/17	IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	603,900	1.00	3,512,959	1.00	0		0		4,116,859	1.00	41	1.00
2011/2012	543,500	0.90	3,125,534	0.89	0		0		3,669,034	0.89	43	1.05
2012/2013	759,000	1.26	3,242,750	0.92	0		0		4,001,750	0.97	42	1.02
2013/2014	728,000	1.21	3,669,500	1.04	0		0		4,397,500	1.07	47	1.15
2014/2015	699,000	1.16	4,002,000	1.14	0		0		4,701,000	1.14	48	1.17
2015/2016	748,500	1.24	4,067,000	1.16	0		0		4,815,500	1.17	50	1.22
2016/2017	646,500	1.07	4,159,500	1.18	0		64,500	1.00	4,870,500	1.18	55	1.34
2017/2018	442,500	0.73	3,581,750	1.02	0		502,750	7.79	4,527,000	1.10	50	1.22

Figure 6a Factor IX units issued between April 2010 & March 2018 - *Severe Haemophilia B only*, all patients with a Scottish postcode

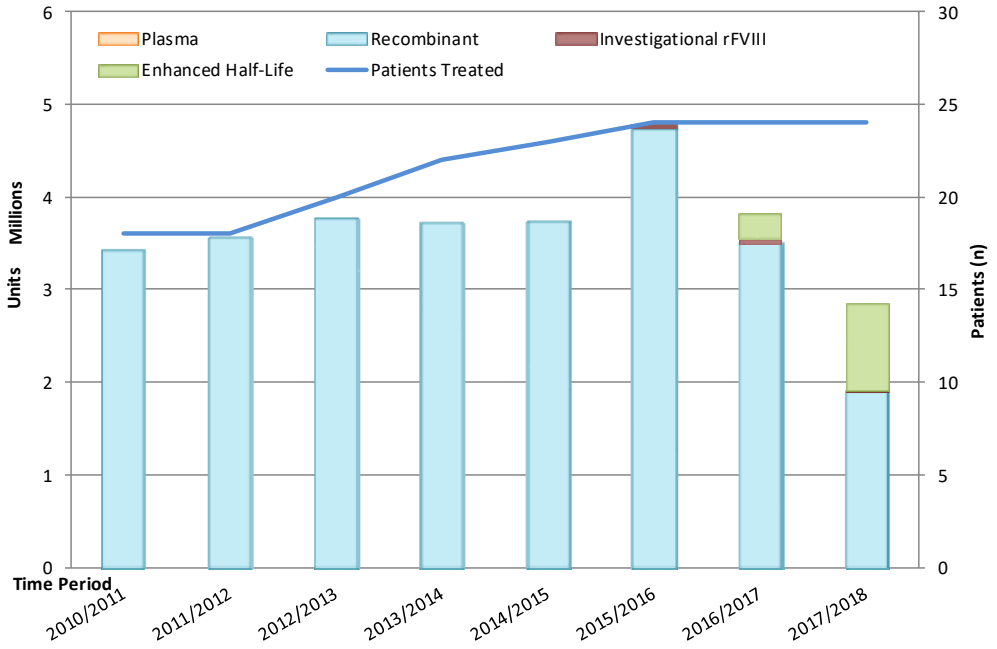


Figure 6b Factor IX units issued between April 2010 & March 2018 - *Severe Haemophilia B only*, patients with a Scotland East postcode

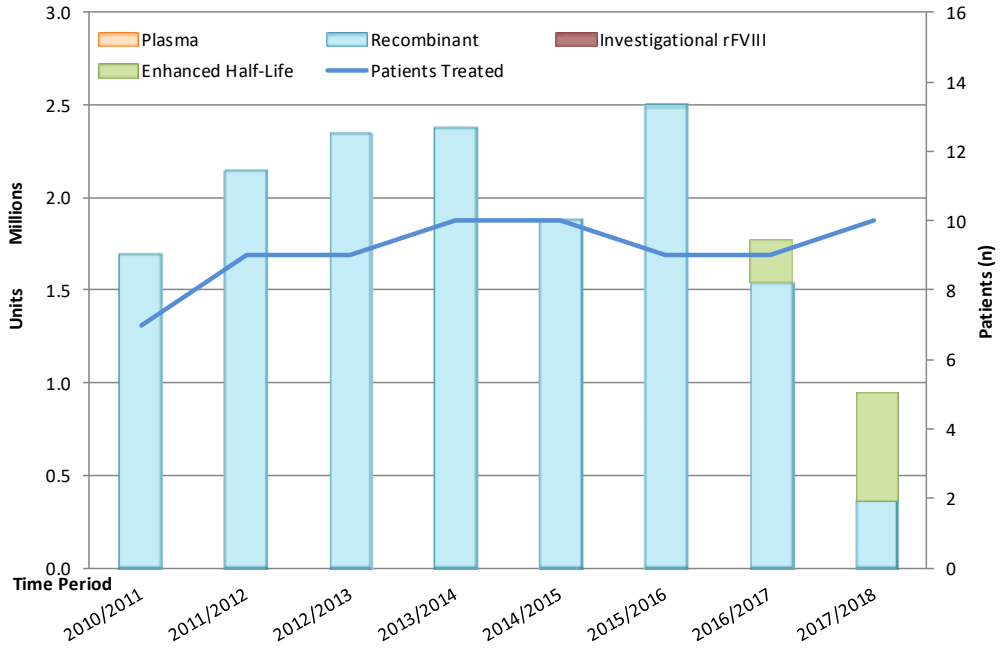
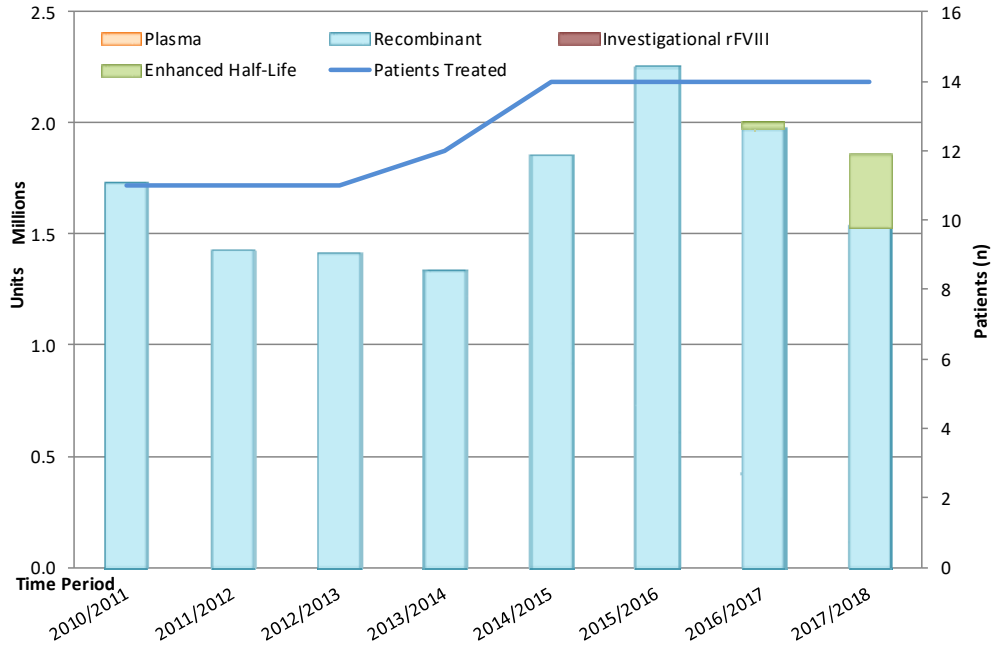


Figure 6c Factor IX units issued between April 2010 & March 2018 – *Severe Haemophilia B only*, patients with a Scotland West postcode



Figures 6a to 6c give an historical view of the number of factor IX units issued between 2010/11 and 2017/18 for patients with *severe haemophilia B only*. Figure 6a includes all patients with a Scottish postcode. Figures 6b includes only patients with a Scotland East postcode and Figure 6c those with a Scotland West postcode. The number of patients 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 patients 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 patients 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 21a Data for Figure 6a - Factor IX units issued between April 2010 & March 2018 - *Severe Haemophilia B*, all Scottish postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU		IU	% difference since 2010/11	IU	% difference since 2015/16	IU	% difference since 2016/17	IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	0		3,427,530	1.00	0		0		3,427,530	1.00	18	1.00
2011/2012	0		3,577,060	1.04	0		0		3,577,060	1.04	18	1.00
2012/2013	0		3,765,357	1.10	0		0		3,765,357	1.10	20	1.11
2013/2014	0		3,712,792	1.08	0		0		3,712,792	1.08	22	1.22
2014/2015	0		3,744,000	1.09	0		0		3,744,000	1.09	23	1.28
2015/2016	0		4,763,250	1.39	2,500	1.00	0		4,765,750	1.39	24	1.33
2016/2017	0		3,512,000	1.02	47,653	19.06	256,250	1.00	3,815,903	1.11	24	1.33
2017/2018	0		1,899,000	0.55	25,588	10.24	916,500	3.58	2,841,088	0.83	24	1.33

Table 21b Data for Figure 6b - Factor IX units issued between April 2010 & March 2018 - *Severe Haemophilia B*, Scotland East postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU		IU	% difference since2010/11	IU		IU	% difference since 2016/17	IU	% difference since2010/11	n	% difference since2010/11
2010/2011	0		1,698,520	1.00	0		0		1,698,520	1.00	7	1.00
2011/2012	0		2,146,060	1.26	0		0		2,146,060	1.26	9	1.29
2012/2013	0		2,350,107	1.38	0		0		2,350,107	1.38	9	1.29
2013/2014	0		2,375,542	1.40	0		0		2,375,542	1.40	10	1.43
2014/2015	0		1,887,000	1.11	0		0		1,887,000	1.11	10	1.43
2015/2016	0		2,502,000	1.47	0		0		2,502,000	1.47	9	1.29
2016/2017	0		1,536,000	0.90	0		230,500	1.00	1,766,500	1.04	9	1.29
2017/2018	0		360,500	0.21	0		585,750	2.54	946,250	0.56	10	1.43

Table 21c Data for Figure 6c - Factor IX units issued between April 2010 & March 2018 - *Severe Haemophilia B*, Scotland West postcodes

Year	Plasma		Recombinant		Investigational rFVIII		Enhanced Half-Life		Total		Patients Treated	
	IU		IU	% difference since2010/11	IU		IU	% difference since 2016/17	IU	% difference since2010/11	n	% difference since2010/11
2010/2011	0		1,729,010	1.00	0		0		1,729,010	1.00	11	1.00
2011/2012	0		1,431,000	0.83	0		0		1,431,000	0.83	11	1.00
2012/2013	0		1,415,250	0.82	0		0		1,415,250	0.82	11	1.00
2013/2014	0		1,337,250	0.77	0		0		1,337,250	0.77	12	1.09
2014/2015	0		1,857,000	1.07	0		0		1,857,000	1.07	14	1.27
2015/2016	0		2,261,250	1.31	0		0		2,261,250	1.31	14	1.27
2016/2017	0		1,976,000	1.14	0		25,750	1.00	2,001,750	1.16	14	1.27
2017/2018	0		1,538,500	0.89	0		322,500	12.52	1,861,000	1.08	14	1.27

Von Willebrand Disease, Rarer Bleeding Disorders and Acquired Defects

Tables 21 - 23 shows the number of patients with Scottish postcodes and reported products issued to treat von Willebrand disease, selected rarer disorders and acquired bleeding disorders during 2017/18, broken down by supplier.

Table 21 Concentrates issued to treat von Willebrand Disease

Manufacturer	Product / Patients (n)	Total Units
CSL Behring	Haemate P (n= 1)	54,500
	Voncento (n= 58)	687,000
Novo Nordisk	NovoSeven (mg) (n= 2)	99
	Desmopressin (n= 43)	792.5

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.XI Deficiency	F.XIII Deficiency
BPL	FXI (n= 2)	-	2,760	-
CSL Behring	Fibrogammin P (n= 3)	-	-	38,250
Novo Nordisk	NovoSeven (mg) (n= 9)	21	-	-
	NovoThirteen (n= 1)	-	-	32,500

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= 4)	-	18,000
Novo Nordisk	NovoSeven (mg) (n= 2)	75	-
Shire	Advate (n= 1)	12,000	-
	FEIBA (n= 3)	136,500	-

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

Table 24 Inhibitors by disease severity

Coagulation Defect	Severity (iu/dl) / Subtype	In Register *	Inhibitors		
			New n (%)	Ongoing n (%)	Historical n (%)
Haemophilia A	< 1	147	1 (0.7)	15 (10.2)	33 (22.4)
	1 - 5	67	0 (0.0)	3 (4.5)	7 (10.4)
	> 5	257	0 (0.0)	0 (0.0)	2 (0.8)
	Total	**471**	**1 (0.2)**	**18 (3.8)**	**42 (8.9)**
Haemophilia B	< 1	24	0 (0.0)	0 (0.0)	0 (0.0)
	1 - 5	43	0 (0.0)	0 (0.0)	0 (0.0)
	> 5	57	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**124**	**0 (0.0)**	**0 (0.0)**	**0 (0.0)**
von Willebrand disease	Type 3	12	0 (0.0)	1 (8.3)	1 (8.3)
	Others	1,141	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**1,162**	**0 (0.0)**	**1 (0.1)**	**1 (0.1)**

** Including patients not regularly treated*

Table 24 shows the incidence of new inhibitors during 2017/18, 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 2017/18. 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 patients with congenital bleeding disorders reported to have a positive inhibitor during 2017/18

Manufacturer	Product / Patients (n)	Units
Haemophilia A		
CSL Behring	Helixate Nexgen (n= 1)	385,500
Novo Nordisk	NovoSeven (mg) (n= 4)	927
Pfizer	ReFacto AF (n= 4)	855,750
Shire	Advate (n= 1)	372,000
	FEIBA (n= 7)	5,253,500
von Willebrand Disease		
Novo Nordisk	NovoSeven (mg) (n= 1)	90
Co-inherited diagnoses		
Novo Nordisk	NovoSeven (mg) (n= 1)	120
Roche	Emicizumab (EAMS) (n= 1)	900
Shire	FEIBA (n= 1)	978,000

Units in IU unless otherwise stated

Table 25 shows the number of patients with Scottish postcodes and reported products issued to patients with an inhibitor newly reported or ongoing during 2017/18, broken down by diagnosis and supplier.

Table 26 Adverse Events

Adverse Event	Number of Events
Allergy Event	0
Infection Event	0
Intracranial haemorrhage	1
Inhibitor Event	1
Malignancy Event	7
Other Event	2
Poor Efficacy Event	0
Thrombotic Event	3
Total	**14**

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 2017/18.

Table 27 Causes of Death

Coagulation Defect	Cause of Death	Severity (factor level iu/dl)			
		< 1	1 - 5	> 5	Total
Haemophilia A	Infection (Bacterial)	0	0	1	**1**
Acquired Haemophilia A	Infection (Bacterial)	1	1	1	**3**
	Senility/Alzheimer's disease	0	1	0	**1**
Haemophilia B	Ruptured Aorta (Peripheral vascular disease)	0	0	1	**1**
	Unknown	0	0	1	**1**
F.VII deficiency	Haemorrhage (Misc)	0	0	1	**1**
F.XI Deficiency	Infection (Bacterial)	0	0	1	**1**
	Ischaemic Heart Disease	0	1	0	**1**
Dysfibrinogenemia	Infection (Bacterial)				**1**
	Renal Failure				**1**
	Total	**1**	**3**	**6**	**12**

Table 28 shows the causes of death reported in patients with a Scottish postcode during 2017/18, broken down by diagnosis and disease severity.