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

Bleeding Disorder Statistics for Scotland

April 2020 to March 2021

A report from the UK National Haemophilia Database

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

Contents

Appendix 1: Glossary

AE	Adverse Event
AGM	Annual General Meeting
ASH	American Society of Hematology
BCSH	British Committee for Standards in Haematology
BMI	Body mass index
BMS	Biomedical Scientists
BSH	British Society for Haematology
CCC	Comprehensive Care Centre
CEO	Chief executive officer
CMWP	Co-morbidities Working Party
COVID-19	Corona Virus Disease
CPD	Continuing Professional Development
CQUIN	Commissioning for Quality and Innovation
CRG	Clinical Reference Group
DAG	Data Analysis Group
DMWP	Data Management Working Party
EAHAD	European Association for Haemophilia and Allied Disorders
EHL	Enhanced Half-life
EU	European Union
EUHASS	European Haemophilia Safety Surveillance
FEIBA	Factor eight inhibitor bypass agent
FIX	Factor nine
FVII	Factor seven
FVIII	Factor eight
GCP	Good clinical practice
GLH	Genomics Laboratory Hub
GLN	Genetic Laboratory Network
GOSH	Great Ormond Street Hospital
GWP	Genetics Working Party
HC	Haemophilia Centre
HCC	Hepatocellular carcinoma
HCIS	Haemophilia Clinical Information System

HCPA	Haemophilia Chartered Physiotherapists' Association
HCV	Hepatitis C virus
HEE	Health Education England
HJHS	Haemophilia Joint Health Score
HNA	Haemophilia Nursing Association
ICS	Integrated Clinical Academic
IPSG	International Prophylaxis Study Group
IQR	Interquartile range
ISTH	International Society on Thrombosis and Haemostasis
ITI	Immune tolerance induction
IU	International units
IU/dl	International units per decilitre
IU/kg	International units per kilogram
IWP	Inhibitor Working Party
kg	Kilogram
MAHA	Microangiopathic hemolytic anemia
MDSAS	Medical Data Solutions and Services
MDT	Multidisciplinary meeting
MTP	Minimally treated patients
NEQAS	National External Quality Assessment Service
NHD	National Haemophilia Database
NHF	National Hemophilia Foundation
NHS	National Health Service
NIBSC	National Institute for Biological Standards and Control
NIHR	National Institute for Health Research
PC	Personal computer
PDF	Portable Document Format
pd-FVIII	Plasma-derived factor eight
pd-FIX	Plasma-derived factor nine
PPIE	Patient and Public Involvement and Engagement
PUP	Previously untreated patient
PwHA	People with haemophilia A
PwHB	People with haemophilia B
PWP	Paediatric Working Party
PwSHA	People with severe haemophilia A

RCEM	Royal College of Emergency Medicine
RCPCH	Royal College of Paediatrics and Child Health
RfPB	NIHR Research for Patient Benefit
rEHL	Recombinant Enhanced Half-Life
rFIX	Recombinant factor IX
rFVIII	Recombinant factor VIII
rSHL	Recombinant Standard Half-Life
SAE	Serious Adverse Event
SHA	Severe Haemophilia A
SHL	Standard Half-life
SOP	Standard operating procedure
TF	Task Force
THS	The Haemophilia Society
UK	United Kingdom
UKHCDO	United Kingdom Haemophilia Centre Doctors' Organisation
UKNEQAS	United Kingdom National External Quality Assessment Service
VWD	Von Willebrand disease
VWF	Von Willebrand factor
WAPPS-Hemo	Web-Accessible Population Pharmacokinetic Service—Hemophilia
WFH	World Federation of Hemophilia
WP	Working party

New Registrations

Table 1 New registrations - Number of people newly registered between April 2020 & March 2021, by diagnosis and gender

Diagnosis	Male	Female	Total
Acquired haemophilia A	4	2	6
Acquired von Willebrand disease	1	1	2
Dysfibrinogenemia	3	4	7
F.VII deficiency	9	11	20
F.X deficiency	2	1	3
F.XI deficiency	6	2	8
Haemophilia A	9	1	10
Haemophilia A carrier	0	1	1
Haemophilia B	2	1	3
Haemophilia B carrier	0	2	2
Hypofibrinogenemia	0	3	3
von Willebrand disease	6	17	23
Other platelet defects	0	1	1
Unclassified bleeding disorder	1	3	4
Miscellaneous	1	0	1
Total	**44**	**50**	**94**

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 2020 & March 2021, by age and disease severity

Diagnosis	Age (years)	Number of people by factor level (IU/dl)		
		< 1	> 5	Total
Haemophilia A	0 - 9	4	2	6
	10 - 19	0	0	0
	20 - 29	0	3	3
	30 - 39	0	0	0
	40 - 49	0	1	1
	50 - 59	0	0	0
	60 - 69	0	0	0
	70 +	0	0	0
	Total	**4**	**6**	**10**
Haemophilia B	0 - 9	0	1	1
	10 - 19	0	1	1
	20 - 29	0	0	0
	30 - 39	0	1	1
	40 - 49	0	0	0
	50 - 59	0	0	0
	60 - 69	0	0	0
	70 +	0	0	0
	Total	**0**	**3**	**3**

N.B Age calculated at mid-year, 30/09/2020

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 2021 and the number treated between April 2020 & March 2021

Diagnosis	In register				Treated (n)	Treated %
	Males	Females	Unknown	Total		
Acquired deficiency (other)	1	0	0	1	0	0.00%
Acquired F.V deficiency	0	1	0	1	0	0.00%
Acquired F.XIII deficiency	0	1	0	1	0	0.00%
Acquired haemophilia A	19	24	0	43	7	16.28%
Acquired von Willebrand disease	10	11	1	22	3	13.64%
Afibrinogenemia	1	0	0	1	1	100.00%
Bernard Soulier	2	4	0	6	0	0.00%
Co-inherited diagnoses	7	15	0	22	2	9.09%
Combined V+VIII deficiency	1	2	0	3	2	66.67%
Dysfibrinogenemia	92	149	0	241	3	1.24%
F.V deficiency	11	21	0	32	0	0.00%
F.VII deficiency	122	153	0	275	6	2.18%
F.X deficiency	17	34	0	51	0	0.00%
F.XI deficiency	102	138	0	240	1	0.42%
F.XIII deficiency	2	3	0	5	4	80.00%
Glanzmann's thrombasthenia	3	8	0	11	3	27.27%
Haemophilia A	493	2	0	495	267	53.94%
Haemophilia A carrier	0	246	0	246	9	3.66%
Haemophilia A with liver transplant	3	0	0	3	0	0.00%
Haemophilia B	131	1	0	132	66	50.00%
Haemophilia B carrier	0	72	0	72	6	8.33%
Haemophilia B with liver transplant	1	0	0	1	0	0.00%
Hypodysfibrinogenemia	6	5	0	11	0	0.00%
Hypofibrinogenemia	12	22	1	35	2	5.71%
Miscellaneous	8	27	0	35	0	0.00%
Other platelet defects	58	148	1	207	3	1.45%
Probable von Willebrand disease	14	36	2	52	0	0.00%
Prothrombin deficiency	1	2	0	3	0	0.00%
Unclassified bleeding disorder	7	69	0	76	2	2.63%
von Willebrand disease	374	755	2	1,131	86	7.60%
Totals	**1,498**	**1,949**	**7**	**3,454**	**473**	

Table 3 shows the total number of active registrations of people with a Scottish postcode and the number who were issued treatment during 2020/21.

Table 4 In Register – The total number of people in the register as of 31st March 2021, by diagnosis and registered Haemophilia Centre

Diagnosis	Aberdeen	Dundee	Edinburgh	Glasgow	Inverness	Total
Acquired deficiency (other)	1	0	0	0	0	1
Acquired F.V deficiency	1	0	0	0	0	1
Acquired F.XIII deficiency	0	1	0	0	0	1
Acquired haemophilia A	17	5	2	20	0	44
Acquired prothrombin deficiency	0	0	0	0	0	0
Acquired von Willebrand disease	10	1	2	9	0	22
Afibrinogenemia	1	0	0	0	0	1
Bernard Soulier	4	1	0	2	0	7
Co-inherited diagnoses	13	1	7	1	0	22
Combined II+VII+IX+X deficiency	0	0	0	0	0	0
Combined V+VIII deficiency	2	0	0	1	0	3
Dysfibrinogenemia	179	5	20	36	1	241
F.V deficiency	23	6	0	3	0	32
F.VII deficiency	126	55	63	30	2	276
F.X deficiency	27	13	5	6	0	51
F.XI deficiency	106	24	48	59	4	241
F.XIII deficiency	2	2	0	1	0	5
Glanzmann's thrombasthenia	2	1	3	5	0	11
Haemophilia A	243	71	42	117	26	499
Haemophilia A carrier	163	28	21	28	6	246
Haemophilia A with liver transplant	0	1	1	1	0	3
Haemophilia B	80	9	9	30	3	131
Haemophilia B carrier	44	2	12	14	0	72
Haemophilia B with liver transplant	0	0	1	0	0	1
Hypodysfibrinogenemia	1	0	0	10	0	11
Hypofibrinogenemia	19	6	0	6	4	35
Miscellaneous	24	2	8	1	0	35
Other platelet defects	95	4	19	89	0	207
Platelet-type pseudo von Willebrand disease	0	0	0	0	0	0
Probable von Willebrand disease	16	1	31	4	0	52
Prothrombin deficiency	1	0	0	2	0	3
Unclassified bleeding disorder	72	1	2	1	0	76
von Willebrand disease	627	158	196	119	37	1137
Total	**1899**	**398**	**492**	**595**	**83**	**3,467**

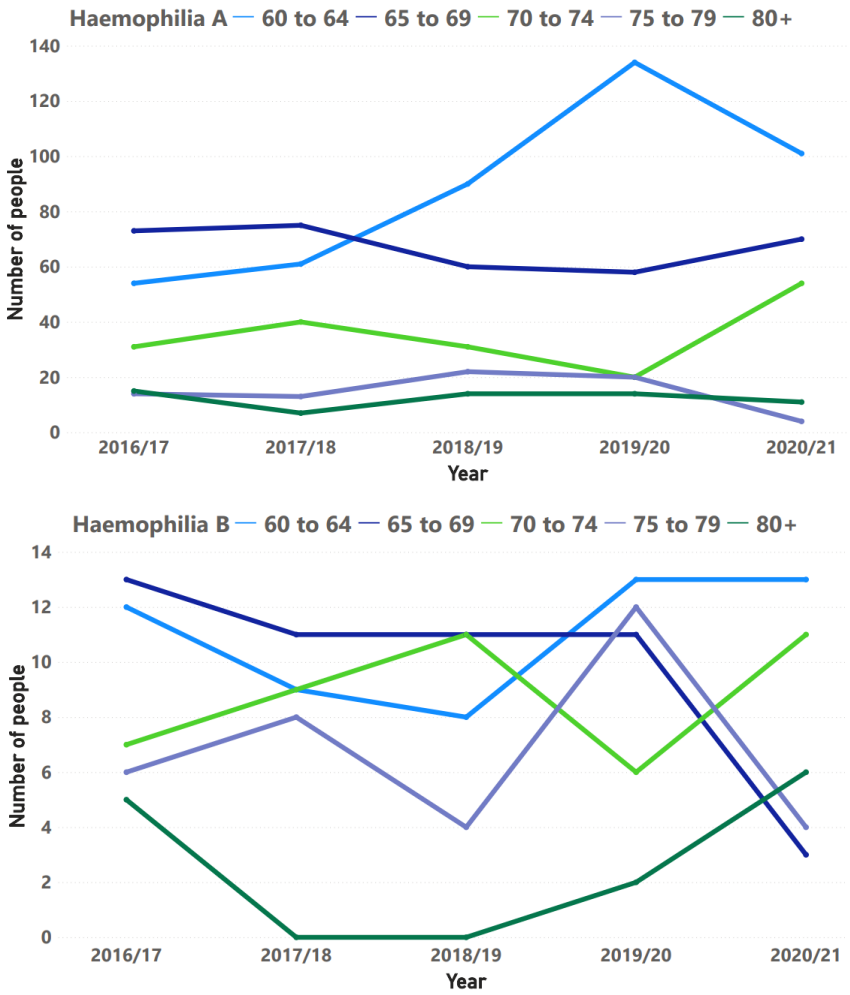
Table 4 shows the number of people registered at each Haemophilia Centre. Patients are allocated to their registered haemophilia centre regardless of their home postcode.

Table 5 In Register – The total number of people with Haemophilia A & B in the register as of 31st March 2021, by severity and age group

Diagnosis	Age (years)	People by factor level (IU/dl)			
		< 1	1 - 5	> 5	Total
Haemophilia A	<18 years	54	13	53	**120**
	≥18 years	97	52	226	**375**
	Total	**151**	**65**	**279**	**495**
Haemophilia B	<18 years	6	7	16	**29**
	≥18 years	18	35	50	**103**
	Total	**24**	**42**	**66**	**132**

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

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



Data tables for figure 1

Year	Age (years)	Number of people by factor VIII level (IU/dl)			
		< 1	1 - 5	> 5	Total
2018/19	60 : 64	50	8	32	90
	65 : 69	43	9	8	60
	70 : 74	8	10	13	31
	75 : 79	5	4	13	22
	80 +	0	4	10	14
2019/20	60 : 64	64	34	36	134
	65 : 69	49	6	3	58
	70 : 74	4	13	3	20
	75 : 79	7	1	12	20
	80 +	0	2	12	14
2020/21	60 : 64	50	18	33	101
	65 : 69	64	4	2	70
	70 : 74	16	32	6	54
	75 : 79	0	2	2	4
	80 +	6	0	5	11

Year	Age (years)	Haemophilia B Number of people by factor IX level (IU/dl)			
		< 1	1 - 5	> 5	Total
2018/19	60 : 64	8	0	0	8
	65 : 69	4	0	7	11
	70 : 74	0	7	4	11
	75 : 79	0	0	3	3
	80 +	0	0	0	0
2019/20	60 : 64	8	0	5	13
	65 : 69	5	1	5	11
	70 : 74	0	5	1	6
	75 : 79	1	0	11	12
	80 +	0	2	0	2
2020/21	60 : 64	10	0	3	13
	65 : 69	0	0	3	3
	70 : 74	4	1	6	11
	75 : 79	0	4	0	4
	80 +	0	0	6	6

Table 6 In Register – The number of people with other selected bleeding disorders in the register as of 31st March 2021 and the number treated between April 2020 & March 2021, by disease severity

Diagnosis	Number of people by factor level (IU/dl)							
	<5		≥5		N/K		Total	
	In reg	Treated	In reg	Treated	In reg	Treated	In reg	Treated
F.V deficiency	1	0	31	0	0	0	32	0
F.VII deficiency	7	0	268	6	0	0	275	6
F.X deficiency	0	0	51	0	0	0	51	0
F.XI deficiency	9	0	231	1	0	0	240	1
Total	**17**	**-**	**581**	**7**	**-**	**-**	**598**	**7**

Diagnosis	<2		2 - <10		10 - <15		N/K		Total	
	In reg	Treated	In reg	Treated	In reg	Treated	In reg	Treated	In reg	Treated
F.XIII deficiency	2	2	2	2	1	0	0	0	5	4
Total	**2**	**2**	**2**	**2**	**1**	**-**	**-**	**-**	**5**	**4**

Table 6 shows the number of people with other selected bleeding disorders and a Scottish postcode known to the NHD during 2020/21. It is acknowledged that these 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 2021 and the number treated between April 2020 & March 2021, 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	14	6	0	**20**	5	34	64	0	**103**	**123**	**6**
Type 2A	2	0	0	0	**2**	2	5	0	0	**7**	**9**	**2**
Type 2B	0	0	0	0	**0**	0	2	1	0	**3**	**3**	**0**
Type 2M	0	1	0	0	**1**	1	3	2	0	**6**	**7**	**0**
Type 2N	0	1	0	0	**1**	0	1	2	0	**3**	**4**	**0**
Type 2 unspecified	1	0	0	0	**1**	4	1	4	0	**9**	**10**	**2**
Type 3	0	1	0	0	**1**	4	2	0	0	**6**	**7**	**5**
Type unreported	5	19	24	0	**48**	11	46	92	0	**149**	**197**	**14**
Other	0	0	1	0	**1**	0	0	0	0	**0**	**1**	**0**
Low VWF	0	1	3	0	**4**	0	0	9	0	**9**	**13**	**1**
										Sub total males	**374**	**30**
Females												
Type 1	0	12	6	0	**18**	4	77	176	0	**257**	**275**	**16**
Type 2A	1	0	0	0	**1**	7	8	1	0	**16**	**17**	**5**
Type 2B	0	1	1	0	**2**	1	4	3	0	**8**	**10**	**3**
Type 2M	1	2	0	0	**3**	9	12	0	0	**21**	**24**	**3**
Type 2N	0	1	0	0	**1**	1	2	1	0	**4**	**5**	**0**
Type 2 unspecified	1	2	0	0	**3**	0	5	1	0	**6**	**9**	**0**
Type 3	1	1	0	0	**2**	1	2	0	0	**3**	**5**	**2**
Type unreported	7	17	17	0	**41**	19	83	225	1	**328**	**369**	**26**
Low VWF	0	1	3	0	**4**	0	0	37	0	**37**	**41**	**1**
										Sub total females	**727**	**58**
Unknown												
Type unreported	2	0	0	0	**2**	0	0	0	0	**0**	**2**	**0**
										Sub total unknown	**2**	**0**
										Grand total - males, females and unknown	**1,103**	**88**

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 of activity levels 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 2020 & March 2021 and region which issued the treatment, by diagnosis, all severities

Diagnosis	Region issuing treatment	People treated (n)
Haemophilia A	London	1
	Scotland East	126
	Scotland West	140
	South Yorkshire & Bassetlaw	1
	Wessex	2
	Sub total	**270**
Haemophilia A carrier	Scotland East	1
	Scotland West	8
	Sub total	**9**
Acquired haemophilia A	Scotland East	4
	Scotland West	3
	Sub total	**7**
Haemophilia B	Scotland East	25
	Scotland West	42
	Sub total	**67**
Haemophilia B carrier	Scotland East	1
	Scotland West	5
	Sub total	**6**
von Willebrand disease	Birmingham & Black Country	2
	Scotland East	48
	Scotland West	36
	Sub total	**86**
Acquired von Willebrand disease	Scotland East	2
	Scotland West	1
	Sub total	**3**
F.VII deficiency	Scotland East	2
	Scotland West	4
	Sub total	**6**
F.XI deficiency	Scotland East	1
	Sub total	**1**
F.XIII deficiency	Scotland East	3
	Scotland West	1
	Sub total	**4**

Continued overleaf....

Table 8 continued…

Diagnosis	Region issuing treatment	People treated (n)
Combined V+VIII deficiency	Scotland East	1
	Scotland West	1
	Sub total	**2**
Co-inherited diagnoses	Scotland East	2
	Sub total	**2**
Afibrinogenemia	Scotland West	1
		1
Dysfibrinogenemia	Scotland East	2
	Scotland West	1
	Sub total	**3**
Hypodysfibrinogenemia	Scotland West	2
	Sub total	**2**
Glanzmann's thrombasthenia	Scotland East	3
	Sub total	**3**
Other platelet defects	Scotland East	3
	Sub total	**3**
Unclassified bleeding disorder	Scotland West	2
	Sub total	**2**
	Grand total	**477**

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

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

Table 9 People with a non-Scottish postcode, registered & treated at a Scottish Haemophilia Centre between April 2020 & March 2021, by diagnosis, all severities

Diagnosis	People's home postcode region	People registered (n)	People treated (n)
Haemophilia A	East Midlands	1	1
	East of England	1	1
	InvalidPostcode	1	-
	London	4	3
	North East	2	-
	North West	3	2
	South Central	4	3
	South East Coast	3	1
	South West	1	-
	Yorkshire and the Humber	2	2
	Sub total	**22**	**13**
Haemophilia A carrier	East of England	1	1
	North West	1	-
	West Midlands	1	-
	Sub total	**3**	**1**
Haemophilia B	East Midlands	2	2
	North West	1	1
	South Central	1	-
	Sub total	**4**	**3**
Haemophilia B carrier	North West	1	-
	Unknown	2	-
	Sub total	**3**	**-**
von Willebrand disease	London	3	-
	North East	5	2
	North West	3	-
	South Central	3	1
	West Midlands	1	-
	Yorkshire and the Humber	2	-
	Miscellaneous	1	-
	Unknown	1	-
	Sub total	**19**	**3**
Acquired von Willebrands disease	South East Coast	1	-
	Sub total	**1**	**-**
F.VII deficiency	North East	1	-
	South Central	1	-
	Unknown	1	-
	Sub total	**3**	**-**
F.XI deficiency	South East Coast	1	-
	South West	2	-
	Sub total	**3**	**-**
F.XIII deficiency	London	1	1
	Sub total	**1**	**1**
Dysfibrinogenemia	East Midlands	1	-
	Unknown	1	-
Hypofibrinogenemia	London	1	-
Other platelet defects	Miscellaneous	1	-
	Sub total	**4**	**-**
	Grand total	**63**	**21**

The people reported in Table 9 were registered at or issued treatment from a Scottish Haemophilia Centre during 2020/21, however, they have a postcode recorded on the NHD which falls outside of Scotland.

Haemophilia A and product use

Table 10 Factor VIII and Emicizumab issued, by diagnosis (all severities)

Diagnosis	People issued FVIII products (n)	FVIII (IU)			People issued Emicizumab (n)	Emicizumab	Total
		Plasma-derived FVIII	Standard half-life	Enhanced half-life			
Haemophilia A	245	246500	38528251	6190250	44	114920	45079921
Haemophilia A carrier	6	0	24000	0	0	0	24000
Acquired haemophilia A	1	0	71500	0	0	0	71500
von Willebrand disease	50	826500	0	0	0	0	826500
Acquired von Willebrands disease	3	48500	0	0	0	0	48500
Co-inherited diagnoses	0	0	0	0	1	4680	4680
Total	**305**	**1,121,500**	**38,623,751**	**6,190,250**	**45**	**119,600**	**46,055,101**

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 2020/21. Also shown are the number of units issued, broken down by diagnosis and product type. No investigational FVIII reported to have been issued.

There are 44 people with Haemophilia A who have been issued with 114,920 IU of Hemlibra and 1 person with Co-inherited diagnosis who has been issued with 4,680 IU of Hemlibra.

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

Manufacturer	Product	People treated (n)	Total units
Grifols	Fanhdi	1	246,500
Novo Nordisk	Esperoct	15	2,363,500
	NovoEight	44	6,843,500
	NovoSeven (mg)	10	488
Octapharma	Nuwiq	1	60,000
Pfizer	ReFacto AF	87	14,951,751
Roche	Hemlibra (mg)	44	114,920
SOBI/Biogen	Elocta	20	3,826,750
Takeda	Advate	112	16,673,000
	FEIBA	1	50,000
Various manufacturers	Desmopressin	7	176
	Other investigational products	2	800

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 and Emicizumab issued to people with *Severe* Haemophilia A (incl. treatment for inhibitors), by issuing Haemophilia Centre

Haemophilia centre issuing treatment	Severe haemophilia A					
	People treated with FVIII (n)	Total FVIII units	Mean usage	People treated with Emicizumab (n)	Total Emicizumab units	Mean usage
Aberdeen	23	5,025,500	218,500	4	4,740	1,185
Dundee	15	4,054,000	270,267	1	3,600	3,600
Edinburgh	28	7,944,500	283,732	13	37,500	2,885
Glasgow	64	14,525,000	226,953	22	58,385	2,654
Inverness	3	489,000	163,000	1	300	300

Table 12a reports the number of people with severe haemophilia A issued treatment during 2020/21 and the number of units of factor VIII and Hemlibra issued. This is broken down by the haemophilia centre which issued the treatment.

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

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

Region issuing treatment	Severe haemophilia A					
	People treated with FVIII (n)	Total FVIII units	Mean usage	People treated with Emicizumab (n)	Total Emicizumab units	Mean usage
Scotland East	68	17,513,000	257,544	18	46,140	2,563
Scotland West	64	14,525,000	226,953	22	58,385	2,654

Table 12b reports the number of people with severe haemophilia A treated and the number of units of factor VIII and Hemlibra issued during 2020/21. This is broken down by region based on the person's registered haemophilia centre.

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

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

Health board	General population	Severe haemophilia A							
		People (n)	FVIII units	Mean usage	FVIII units per capita	People (n)	Emicizumab units	Mean usage	Emicizumab units per capita
Borders	115,240	2	1,787,000	893,500	15.51	1	1,200	1,200	0.01
Dumfries and Galloway	148,290	4	1,512,501	378,125	10.20	1	4,200	4,200	0.03
Western Isles	26,500	1	180,000	180,000	6.79	0	0	0	0.00
Forth Valley	305,930	6	1,288,500	214,750	4.21	3	12,000	4,000	0.04
Lothian	912,490	18	4,588,500	254,917	5.03	9	20,820	2,313	0.02
Grampian	585,550	20	4,725,750	236,288	8.07	4	11,820	2,955	0.02
Tayside	416,550	10	3,078,000	307,800	7.39	1	3,600	3,600	0.01
Lanarkshire	661,960	6	1,829,500	304,917	2.76	1	6,060	6,060	0.01
Ayrshire and Arran	367,990	11	2,311,250	210,114	6.28	3	10,110	3,370	0.03
Greater Glasgow and Clyde	1,185,240	37	7,909,250	213,764	6.67	14	29,915	2,137	0.03
Highland	320,860	6	1,359,000	226,500	4.24	2	3,360	1,680	0.01
Shetland	22,870	3	443,750	147,917	19.40	0	0	0	0.00
Fife	374,130	9	1,636,500	181,833	4.37	1	1,440	1,440	0.00
Scotland	**5,443,600**	**133**	**32,649,501**	**245,485**	**6.00**	**40**	**104,525**	**2,613**	**0.02**

Ranked by mean usage

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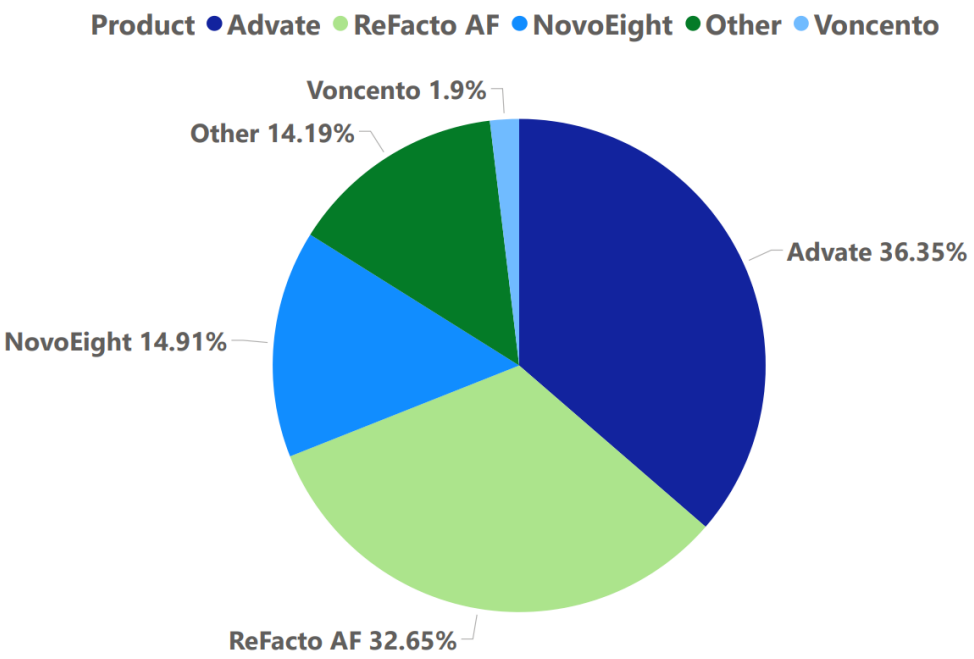
Table 13 reports the number of people with severe haemophilia A issued treatment and number of units of factor VIII issued broken down by Health Board and ranked by the mean number of units issued per patient. Usage per capita of population is also reported.

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

Figure 2 Market share for Factor VIIII concentrates with Haemophilia A between April 2020 & March 2021



Includes products containing a combination of VWF and FVIII, which are reported in FVIII units (IU).

This pie chart does not contain the Hemlibra (mg) units issued.

Manufacturer	Product	Units (IU)	People treated (n)
Takeda	Advate	16,721,000	114
Pfizer	ReFacto AF	14,968,251	91
CSL Behring	Voncento	875,000	53
Roche	Hemlibra (mg)	119,600	45
Novo Nordisk	NovoEight	6,849,500	45
Other	Other	6,521,750	38
	Total	**46,055,101**	**386**

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

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

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

Figure 3a Factor VIII units and Emicizumab issued between April 2013 & March 2021 - all diagnoses, all severities, all people (issued FVIII & any product) with a Scottish postcode

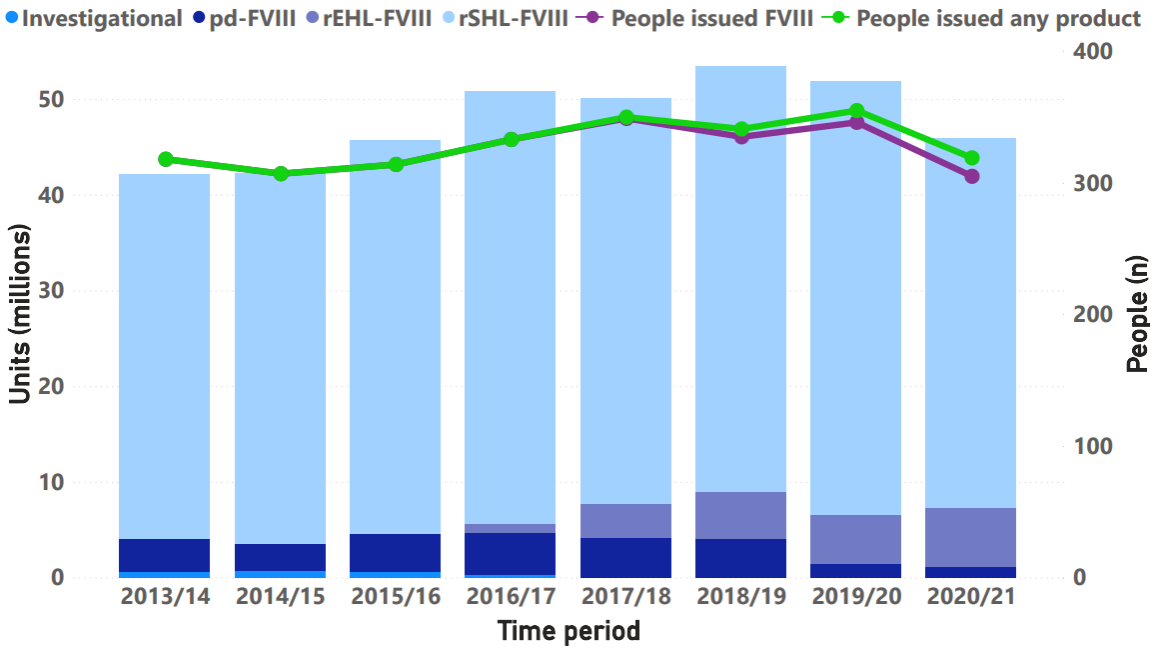


Figure 3b Factor VIII units issued between April 2013 & March 2021 - all diagnoses, all severities, people with a Scotland East postcode

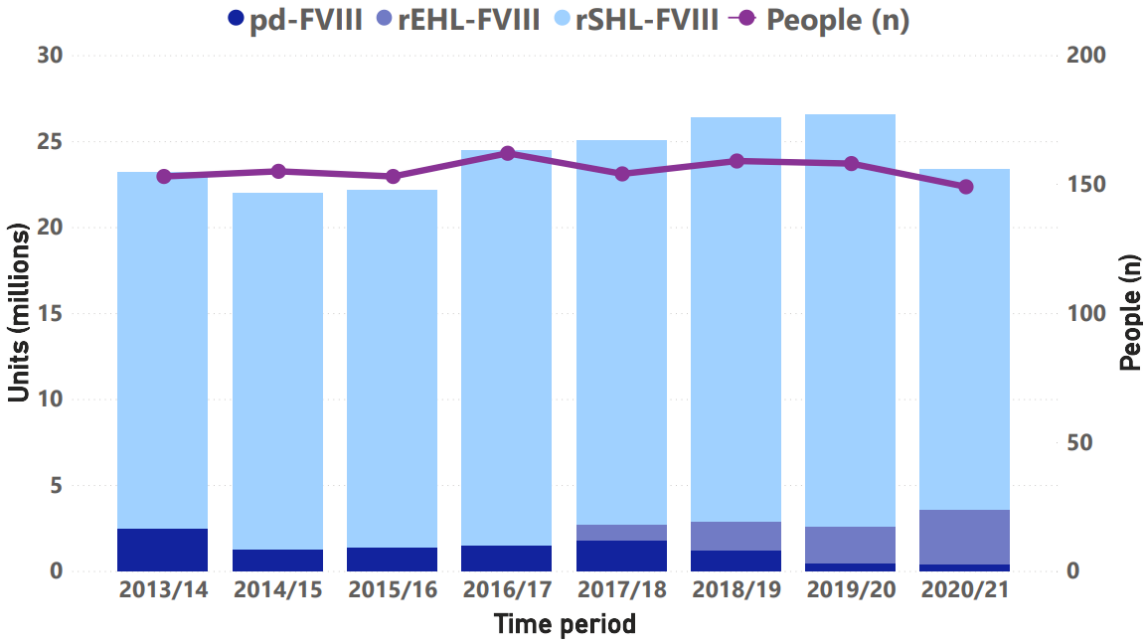
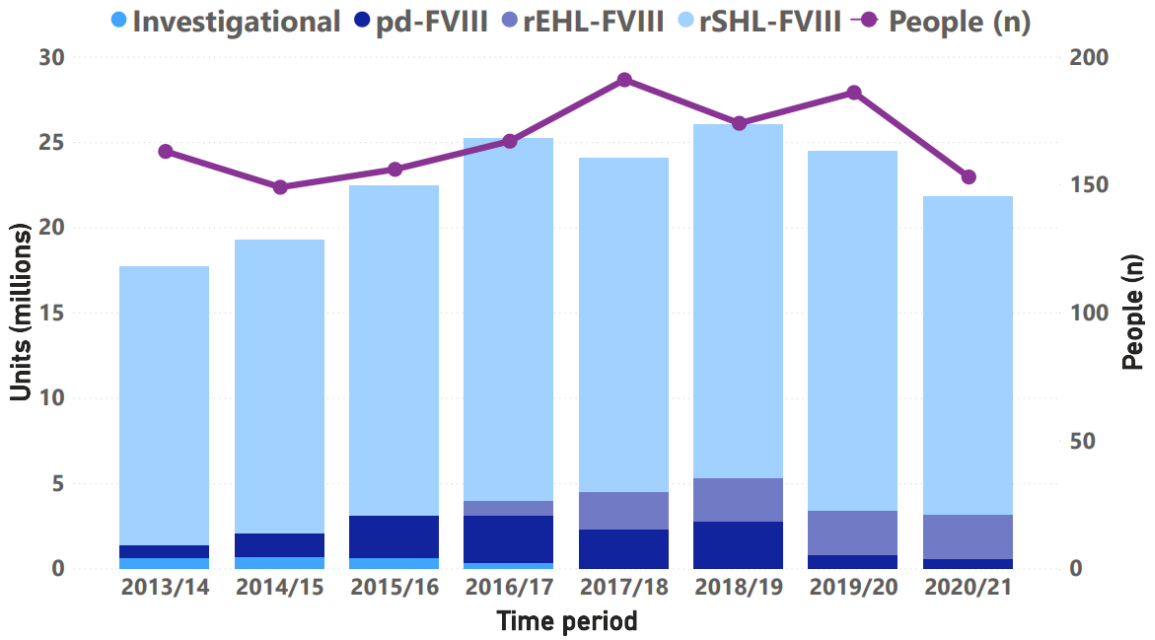


Figure 3c Factor VIII units issued between April 2013 & March 2021 – all diagnoses, all severities, people with a Scotland West postcode



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

Figures 3a to 3c give a historical view of the number of factor VIII units issued between 2013/14 and 2020/21 for all diagnoses and all severities. Figure 3a includes all people with a Scottish postcode. Figures 3b only includes people with a Scotland East postcode and Figure 3c those with a Scotland West postcode. The number of people treated is represented by the green 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.

The apparent fall-off in patients treated with Factor VIII is attributable to patients with severe haemophilia changing to Hemlibra (emicizumab).

Table 14a Data for Figure 3a - Factor VIII and Emicizumab units issued between April 2013 & March 2021 – all diagnoses, all Scottish postcodes

Year	Plasma-derived		Investigational		Recombinant				Total		People issued FVIII		People issued Emicizumab	
					Standard half-life		Enhanced half-life							
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year	n	% difference year on year
2013/2014	3,385,600	-	630,000	-	38,129,645	-	-	-	42,145,245	-	318	-	-	-
2014/2015	2,819,500	-16.72	674,750	+7.1	38,770,500	+1.7	-	-	42,264,750	+0.3	307	-3.46	-	-
2015/2016	3,983,050	+41.3	613,500	-9.08	41,102,750	+6	-	-	45,699,300	+8.1	314	+2.3	-	-
2016/2017	4,402,000	+10.5	330,500	-46.13	45,211,000	+10	849,750	-	50,793,250	+11.1	333	+6.1	-	-
2017/2018	4,222,000	-4.09	-	-	42,394,250	-6.23	3,470,750	-	50,087,000	-1.39	349	+4.8	1	-
2018/2019	4,078,500	-3.40	-	-	44,469,500	+4.9	4,849,950	+39.7	53,397,950	+6.6	335	-4.01	8	+700
2019/2020	1,396,000	-65.77	-	-	45,295,100	+1.9	5,205,500	+7.3	51,896,600	-2.81	346	+3.3	17	+112.5
2020/2021	1,121,500	-19.66	-	-	38,623,751	-14.73	6,190,250	+18.9	45,935,501	-11.49	305	-11.85	45	+164.7

Table 14b Data for Figure 3b - Factor VIII units issued between April 2013 & March 2021 – all diagnoses, Scotland East postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	2,500,600	-	20,722,895	-	-	-	23,223,495	-	153	-
2014/2015	1,273,000	-49.09	20,711,750	-0.05	-	-	21,984,750	-5.33	155	+1.3
2015/2016	1,389,500	+9.2	20,782,250	+0.3	-	-	22,171,750	+0.9	153	-1.29
2016/2017	1,478,500	+6.4	23,005,000	+10.7	-	-	24,483,500	+10.4	162	+5.9
2017/2018	1,786,500	+20.8	22,355,750	-2.82	919,000	-	25,061,250	+2.4	154	-4.94
2018/2019	1,186,000	-33.61	23,484,750	+5.1	1,720,000	+87.2	26,390,750	+5.3	159	+3.2
2019/2020	465,500	-60.75	23,953,250	+2	2,109,500	+22.6	26,528,250	+0.5	158	-0.63
2020/2021	405,500	-12.89	19,771,500	-17.46	3,171,000	+50.3	23,348,000	-11.99	149	-5.70

Table 14c Data for Figure 3c - Factor VIII units issued between April 2013 & March 2021 - all diagnoses, Scotland West postcodes

Year	Plasma-derived		Investigational		Recombinant				Total		People treated	
					Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	729,000	-	630,000	-	16,318,250	-	-	-	17,677,250	-	163	-
2014/2015	1,385,500	+90.1	674,750	+7.1	17,197,250	+5.4	-	-	19,257,500	+8.9	149	-8.59
2015/2016	2,470,050	+78.3	613,500	-9.08	19,358,000	+12.6	-	-	22,441,550	+16.5	156	+4.7
2016/2017	2,785,500	+12.8	330,500	-46.13	21,225,250	+9.6	849,750	-	25,191,000	+12.3	167	+7.1
2017/2018	2,319,500	-16.73	-	-	19,582,500	-7.74	2,170,750	+155.5	24,072,750	-4.44	191	+14.4
2018/2019	2,748,500	+18.5	-	-	20,708,500	+5.8	2,580,950	+18.9	26,037,950	+8.2	174	-8.90
2019/2020	779,500	-71.64	-	-	21,075,100	+1.8	2,601,000	+0.8	24,455,600	-6.08	186	+6.9
2020/2021	574,500	-26.30	-	-	18,660,000	-11.46	2,599,250	-0.07	21,833,750	-10.72	153	-17.74

Figure 4a Factor VIII units issued between April 2013 & March 2021 - *Severe Haemophilia A only*, all people (issued FVIII & any product) with a Scottish postcode

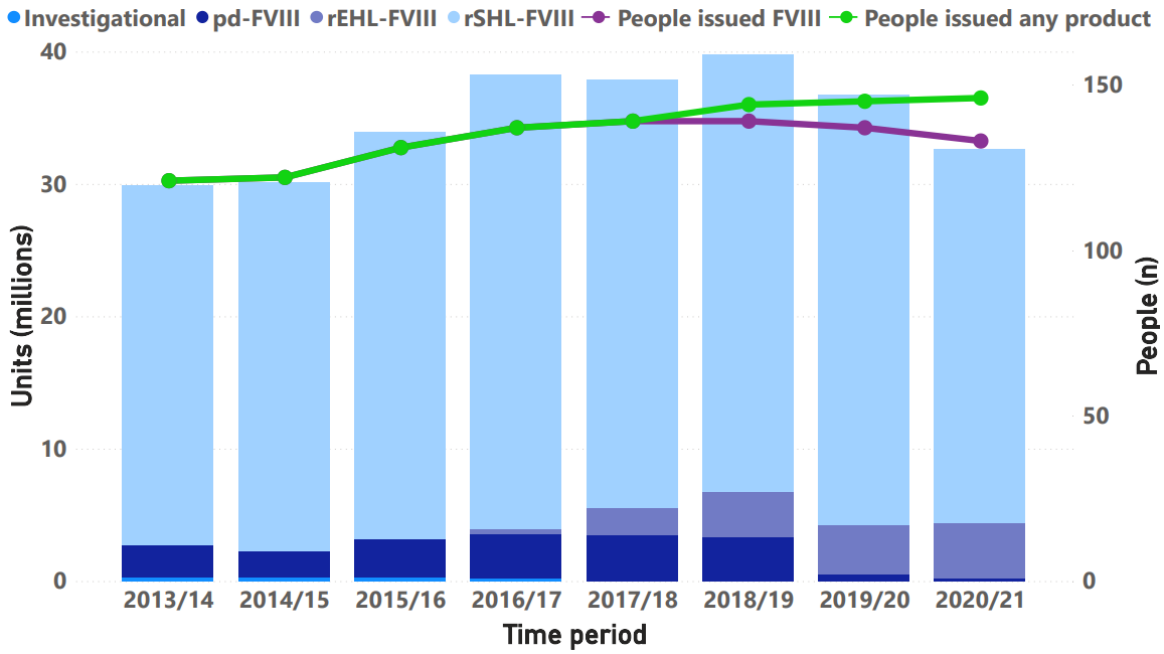


Figure 4b Factor VIII units issued between April 2013 & March 2021 - *Severe Haemophilia A only*, people with a Scotland East postcode

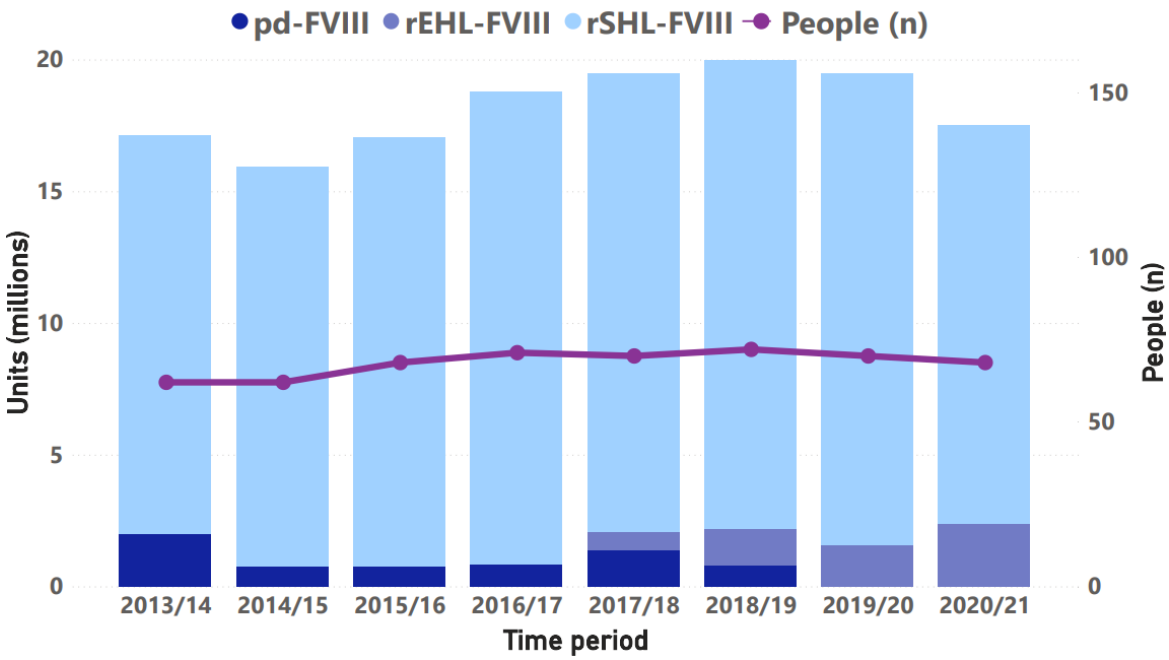
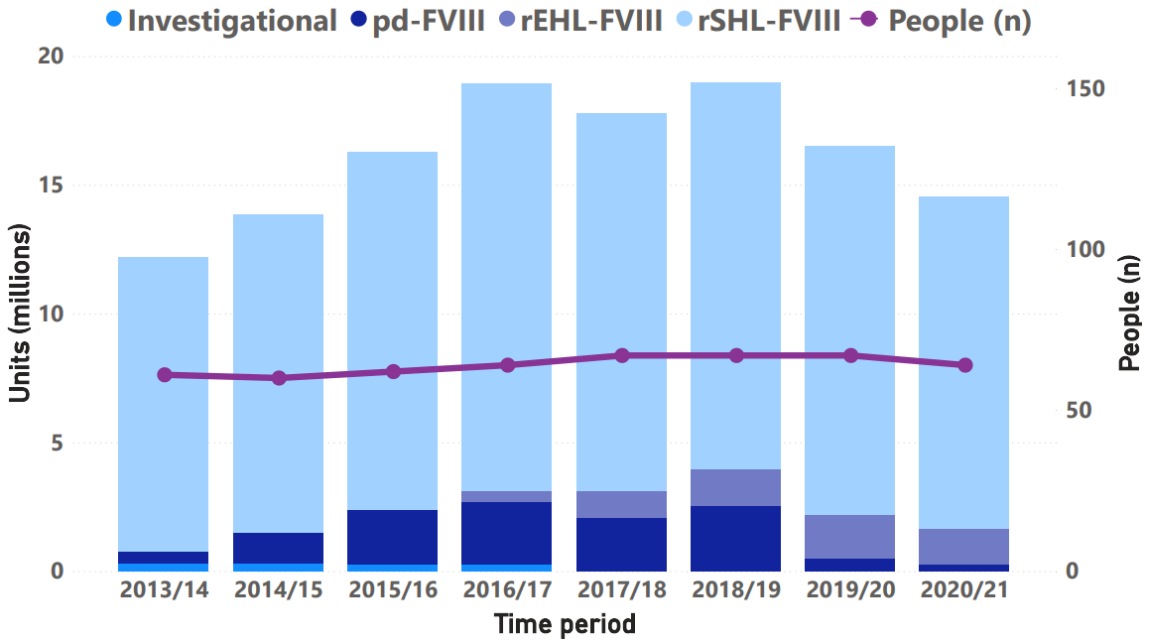


Figure 4c Factor VIII units issued between April 2013 & March 2021 – *Severe Haemophilia A only*, people with a Scotland West postcode



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

Figures 4a to 4c give a historical view of the number of factor VIII units issued between 2013/14 and 2020/21 for people with *severe haemophilia A only*. Figure 4a includes all patients with a Scottish postcode. Figures 4b includes only people with a Scotland East postcode and Figure 4c those with a Scotland West postcode. The number of people treated is represented by the green line using a secondary axis.

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

NOTE: Figure 4a is based on all people with a Scottish postcode regardless of which haemophilia centre issued the product. For Figures 4b and 4c the location of the haemophilia centre is used to identify the geographical region. This means that Figure 4a is not directly comparable to Figures 4b and 4c as some people are dropped due to the fact that they were issued treatment from a haemophilia centre outside of Scotland. In addition, some people were issued treatment from both East and West Scotland. The fall-off in the amount of Factor VIII used is attributable to switching of patients with severe haemophilia to Hemlibra (emicizumab).

Table 15a Data for Figure 4a - Factor VIII units issued between April 2013 & March 2021 - *Severe Haemophilia A*, all Scottish postcodes

Year	Plasma-derived		Investigational		Recombinant				Total		People issued FVIII		People issued Emicizumab	
					Standard half-life		Enhanced half-life							
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year	n	% difference year on year
2013/2014	2,456,000	-	293,000	-	27,199,355	-	-	-	29,948,355	-	121	-	-	-
2014/2015	1,923,000	-21.70	314,750	+7.4	27,885,750	+2.5	-	-	30,123,500	+0.6	122	+0.8	-	-
2015/2016	2,882,500	+49.9	253,500	-19.46	30,770,250	+10.3	-	-	33,906,250	+12.6	131	+7.4	-	-
2016/2017	3,295,500	+14.3	240,500	-5.13	34,319,250	+11.5	430,750	-	38,286,000	+12.9	137	+4.6	-	-
2017/2018	3,459,000	+5	-	-	32,368,750	-5.68	2,097,250	+386.9	37,925,000	-0.94	139	+1.5	-	-
2018/2019	3,340,000	-3.44	-	-	33,020,250	+2	3,378,200	+61.1	39,738,450	+4.8	139	-	7	-
2019/2020	485,500	-85.46	-	-	32,484,350	-1.62	3,766,500	+11.5	36,736,350	-7.55	137	-1.44	16	+128.6
2020/2021	246,500	-49.23	-	-	28,203,251	-13.18	4,199,750	+11.5	32,649,501	-11.12	133	-2.92	40	+150

Table 15b Data for Figure 4b - Factor VIII units issued between April 2013 & March 2021 - *Severe Haemophilia A*, Scotland East postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	1,988,000	-	15,129,605	-	-	-	17,117,605	-	62	-
2014/2015	754,000	-62.07	15,157,250	+0.2	-	-	15,911,250	-7.05	62	-
2015/2016	763,000	+1.2	16,283,750	+7.4	-	-	17,046,750	+7.1	68	+9.7
2016/2017	834,000	+9.3	17,951,000	+10.2	-	-	18,785,000	+10.2	71	+4.4
2017/2018	1,396,000	+67.4	17,406,500	-3.03	662,000	-	19,464,500	+3.6	70	-1.41
2018/2019	806,500	-42.23	17,793,000	+2.2	1,386,000	+109.4	19,985,500	+2.7	72	+2.9
2019/2020	-	-	17,898,750	+0.6	1,580,500	+14	19,479,250	-2.53	70	-2.78
2020/2021	-	-	15,139,500	-15.42	2,373,500	+50.2	17,513,000	-10.09	68	-2.86

Table 15c Data for Figure 4c - Factor VIII units issued between April 2013 & March 2021 - *Severe Haemophilia A*, Scotland West postcodes

Year	Plasma-derived		Investigational		Recombinant				Total		People treated	
					Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	468,000	-	293,000	-	11,409,750	-	-	-	12,170,750	-	61	-
2014/2015	1,169,000	+149.8	314,750	+7.4	12,353,500	+8.3	-	-	13,837,250	+13.7	60	-1.64
2015/2016	2,119,500	+81.3	253,500	-19.46	13,876,000	+12.3	-	-	16,249,000	+17.4	62	+3.3
2016/2017	2,461,500	+16.1	240,500	-5.13	15,804,500	+13.9	430,750	-	18,937,250	+16.5	64	+3.2
2017/2018	2,063,000	-16.19	-	-	14,667,750	-7.19	1,054,250	+144.7	17,785,000	-6.08	67	+4.7
2018/2019	2,533,500	+22.8	-	-	15,017,250	+2.4	1,443,200	+36.9	18,993,950	+6.8	67	-
2019/2020	485,500	-80.84	-	-	14,323,100	-4.62	1,691,000	+17.2	16,499,600	-13.13	67	-
2020/2021	246,500	-49.23	-	-	12,872,250	-10.13	1,406,250	-16.84	14,525,000	-11.97	64	-4.48

Haemophilia B and Factor IX use

Table 16 Factor IX issued, by diagnosis

Diagnosis	People treated (n)	FIX (IU)		
		Recombinant	Enhanced half-life	Total
Haemophilia B	65	4,092,750	2,651,000	6,743,750
Haemophilia B carrier	6	30,000	-	30,000
Total	**71**	**4,122,750**	**2,651,000**	**6,773,750**

Table 16 shows the number of people with a Scottish postcode who were issued factor IX concentrate during 2020/21. 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	People treated (n)	Total units
CSL Behring	IDELVION	21	2,100,750
Novo Nordisk	NovoSeven (mg)	1	32
	Refixia	2	112,000
Pfizer	Benefix	41	4,092,750
SOBI/Biogen	ALPROLIX	3	438,250
Various manufacturers	Other investigational products	1	480

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		
	People treated (n)	Total FIX units	Mean usage
Aberdeen	4	334,500	83,625
Dundee	5	402,750	80,550
Glasgow	13	2,137,750	164,442

Table 18 reports the number of people with severe haemophilia B treated and the number of units of factor IX issued during 2020/21. This is broken down by the haemophilia centre which issued the treatment. Comparisons between centres for mean usage per patient are inappropriate for haemophilia B, given large interpersonal variation and the very small number of patients involved.

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

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

Region issuing treatment	Severe haemophilia B		
	People treated (n)	Total FIX units	Mean usage
Scotland East	9	737,250	81,917
Scotland West	13	2,137,750	164,442
	22	**2,875,000**	**130,682**

Table 18b reports the number of people with severe haemophilia B treated and the number of units of factor IX issued during 2020/21. This is broken down by region based on the person's postcode as recorded on the NHD.

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

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

Health board	General population	Severe haemophilia B			
		People (n)	FIX units	Mean usage	FIX units per capita
Ayrshire and Arran	115,240	2	391,500	195,750	3.40
Forth Valley	305,930	1	512,000	512,000	1.67
Grampian	585,550	4	334,500	83,625	0.57
Greater Glasgow and Clyde	1,185,240	5	628,000	125,600	0.53
Highland	320,860	1	168,000	168,000	0.52
Lanarkshire	661,960	4	438,250	109,563	0.66
Tayside	416,550	5	402,750	80,550	0.97
Scotland	**3,591,330**	**22**	**2,875,000**	**130,682**	**0.80**

Ranked by mean usage

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

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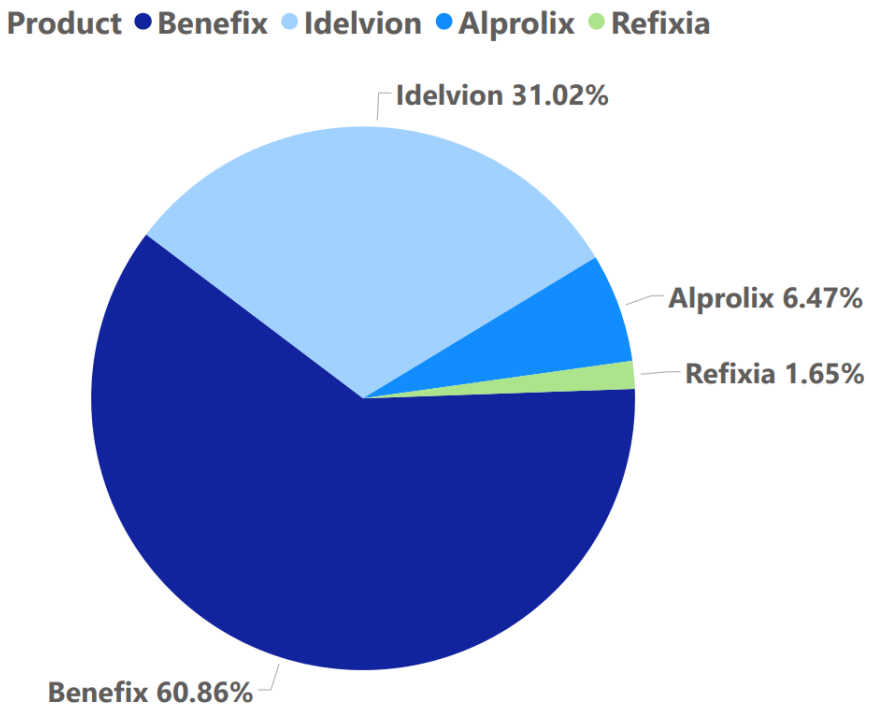
Table 19 reports the number of people with severe haemophilia B treated and number of units of factor IX issued broken down by Health Board and ranked by the mean number of units issued per person. Usage per capita of population is also reported.

This table does not contain duplicate numbers of people. People are allocated to a Health Board based on their home postcode. It is recognised that factor IX requirement will vary considerably between individuals.

Figure 5 Market share of Factor IX concentrates issued to people with a Scottish postcode between April 2020 & March 2021



Manufacturer	Product	Units (IU)	People treated (n)
Pfizer	Benefix	4,122,750	47
CSL Behring	Idelvion	2,100,750	21
SOBI/Biogen	Alprolix	438,250	3
NovoNordisk	Refixia	112,000	2
	Total	**6,773,750**	**73**

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

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

Figure 6a Factor IX units issued between April 2013 & March 2021 - all diagnoses, all severities, all people with a Scottish postcode

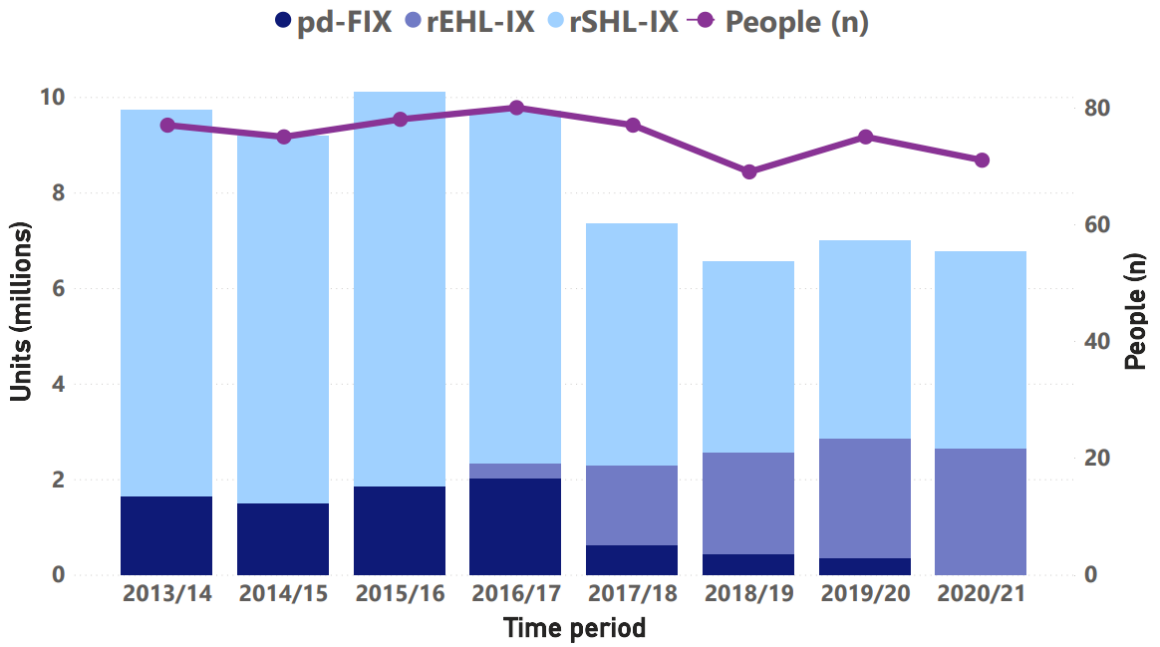


Figure 6b Factor IX units issued between April 2013 & March 2021 - all diagnoses, all severities, people with a Scotland East postcode

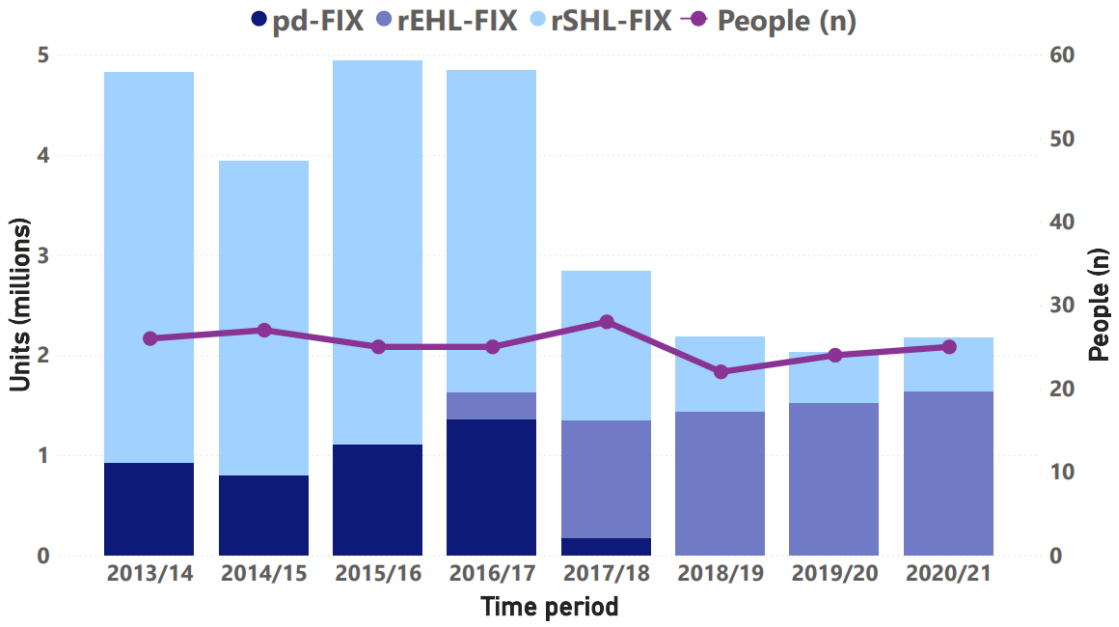
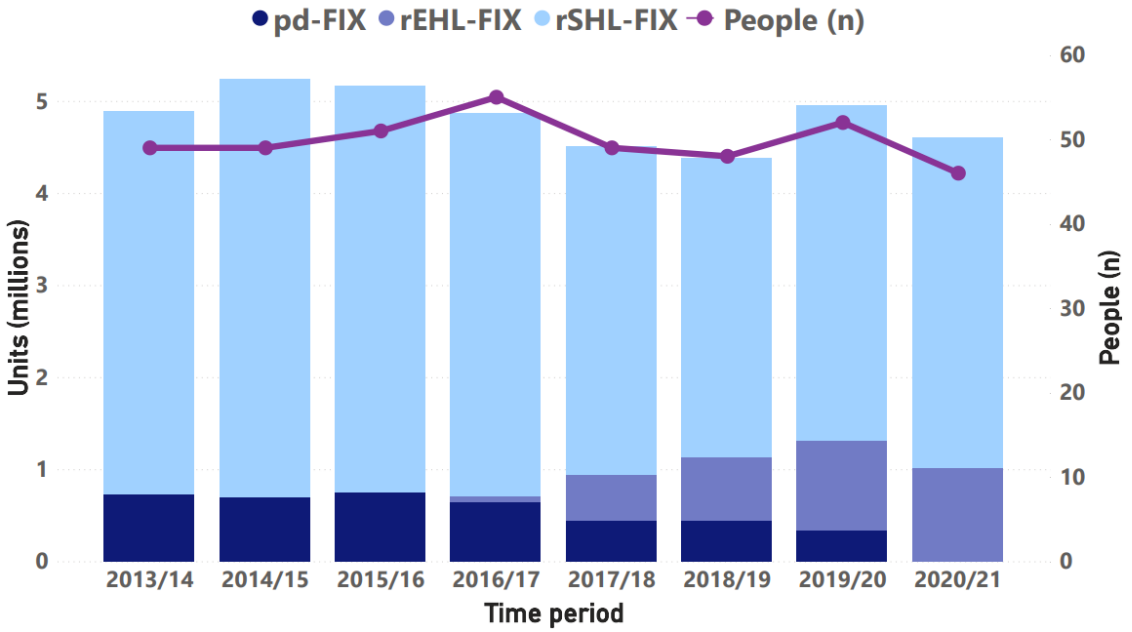


Figure 6c Factor IX units issued between April 2013 & March 2021 – all diagnoses, all severities, people with a Scotland West postcode



Figures 6a to 6c give a historical view of the number of factor IX units issued between 2013/14 and 2020/21 for all diagnoses and all severities. Figure 6a includes all people with a Scottish postcode. Figures 6b includes only people with a Scotland East postcode and Figure 6c those with a Scotland West postcode. The number of people treated is represented by the green line using a secondary axis.

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

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

The apparent fall off in number of factor IX units used is attributable to a change from standard half-life to extended half-life factor IX products. Fewer factor IX units are required when extended half-life products are used because of the longer half-life of the product. This change in clinical practice is also associated with a dramatic reduction in the use of plasma-derived factor VIII.

Table 20a Data for figure 6a - Factor IX units issued between April 2013 & March 2021 - all diagnoses, all Scottish postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	1,656,935	-	8,079,089	-	-	-	9,736,024	-	77	-
2014/2015	1,501,780	-9.36	7,685,500	-4.87	-	-	9,187,280	-5.64	75	-2.60
2015/2016	1,862,370	+24	8,253,000	+7.4	-	-	10,115,370	+10.1	78	+4
2016/2017	2,013,200	+8.1	7,381,000	-10.57	325,500	-	9,719,700	-3.91	80	+2.6
2017/2018	614,305	-69.49	5,056,250	-31.50	1,684,250	+417.4	7,354,805	-24.33	77	-3.75
2018/2019	441,000	-28.21	3,999,250	-20.90	2,130,250	+26.5	6,570,500	-10.66	69	-10.39
2019/2020	340,000	-22.90	4,145,250	+3.7	2,503,500	+17.5	6,988,750	+6.4	75	+8.7
2020/2021	-	-	4,122,750	-0.54	2,651,000	+5.9	6,773,750	-3.08	71	-5.33

Table 20b Data for figure 6b - Factor IX units issued between April 2013 & March 2021 - all diagnoses, Scotland East postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	928,935	-	3,906,589	-	-	-	4,835,524	-	26	-
2014/2015	802,780	-13.58	3,143,500	-19.53	-	-	3,946,280	-18.39	27	+3.8
2015/2016	1,113,870	+38.8	3,831,000	+21.9	-	-	4,944,870	+25.3	25	-7.41
2016/2017	1,366,700	+22.7	3,221,500	-15.91	261,000	-	4,849,200	-1.93	25	-
2017/2018	171,805	-87.43	1,487,000	-53.84	1,181,500	+352.7	2,840,305	-41.43	28	+12
2018/2019	-	-	748,750	-49.65	1,436,000	+21.5	2,184,750	-23.08	22	-21.43
2019/2020	-	-	505,500	-32.49	1,527,500	+6.4	2,033,000	-6.95	24	+9.1
2020/2021	-	-	532,000	+5.2	1,641,250	+7.4	2,173,250	+6.9	25	+4.2

Table 20c Data for Figure 6c - Factor IX units issued between April 2013 & March 2021 - all diagnoses, Scotland West postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	728,000	-	4,159,500	-	-	-	4,887,500	-	49	-
2014/2015	699,000	-3.98	4,542,000	+9.2	-	-	5,241,000	+7.2	49	-
2015/2016	748,500	+7.1	4,411,000	-2.88	-	-	5,159,500	-1.56	51	+4.1
2016/2017	646,500	-13.63	4,159,500	-5.70	64,500	-	4,870,500	-5.60	55	+7.8
2017/2018	442,500	-31.55	3,563,250	-14.33	502,750	-38.77	4,508,500	-7.43	49	-10.91
2018/2019	441,000	-0.34	3,250,500	-8.78	694,250	+38.1	4,385,750	-2.72	48	-2.04
2019/2020	340,000	-22.90	3,639,750	+12	976,000	+40.6	4,955,750	+13	52	+8.3
2020/2021	-	-	3,590,750	-1.35	1,009,750	+3.5	4,600,500	-7.17	46	-11.54

Figure 7a Factor IX units issued between April 2013 & March 2021 - *Severe Haemophilia B only*, all people with a Scottish postcode

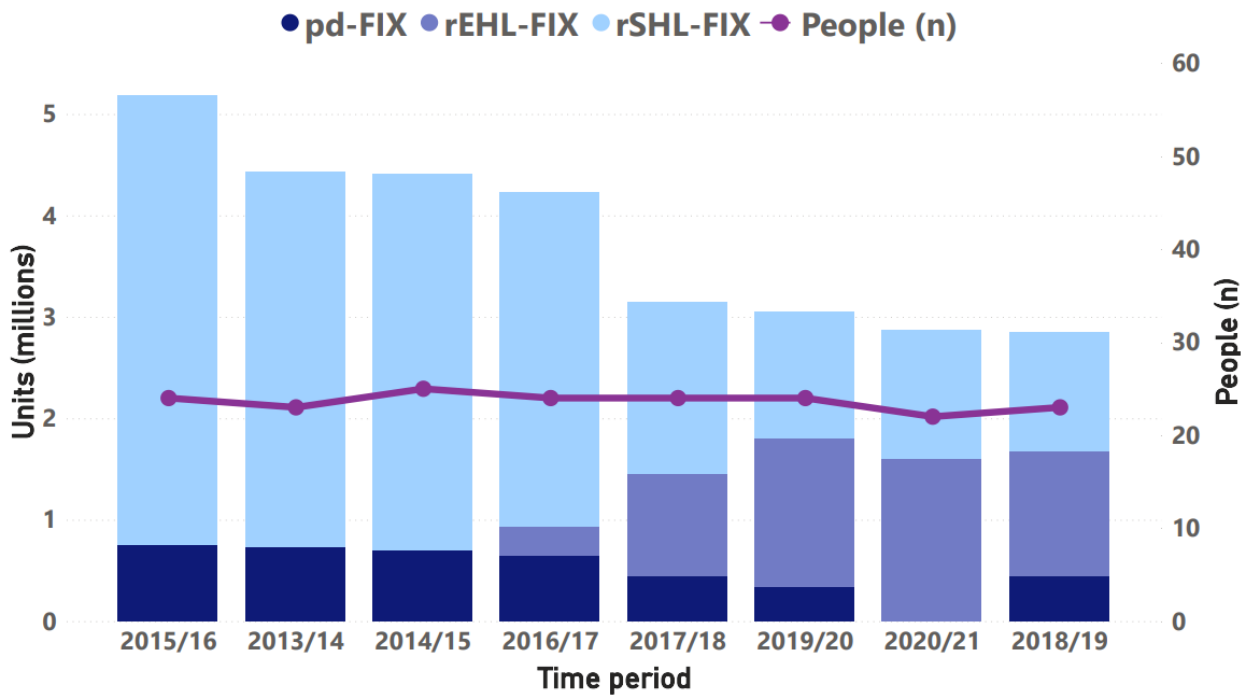


Figure 7b Factor IX units issued between April 2013 & March 2021 - *Severe Haemophilia B only*, people with a Scotland East postcode

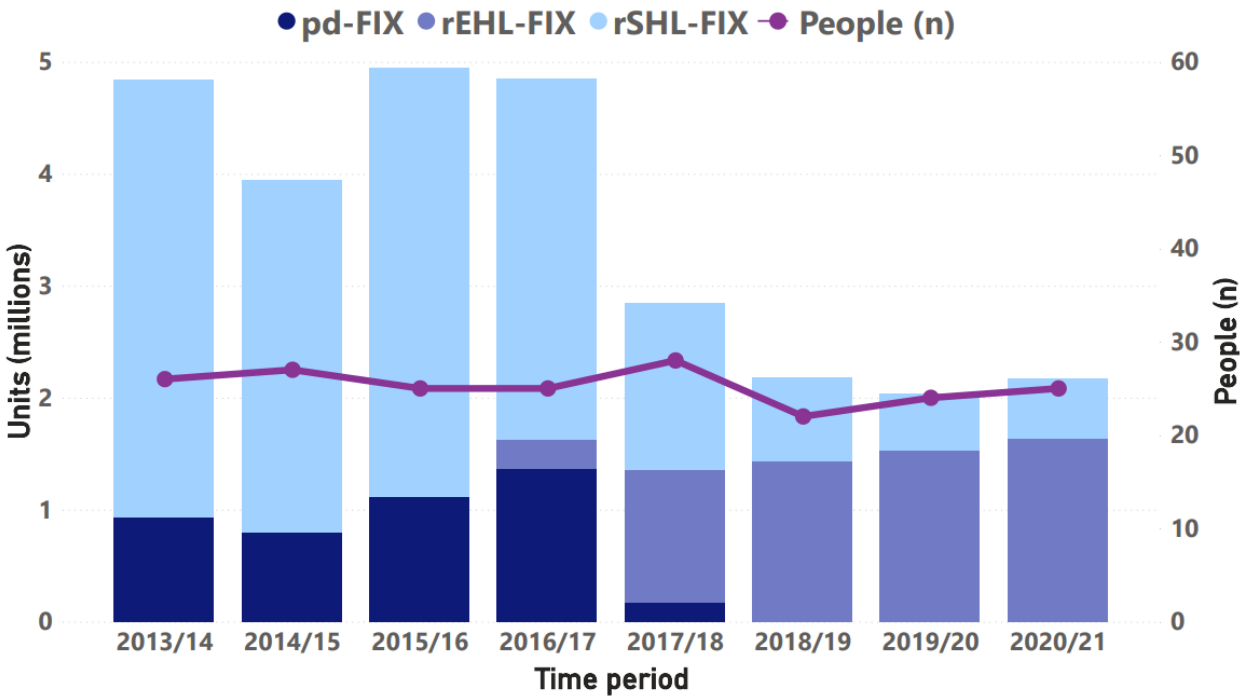
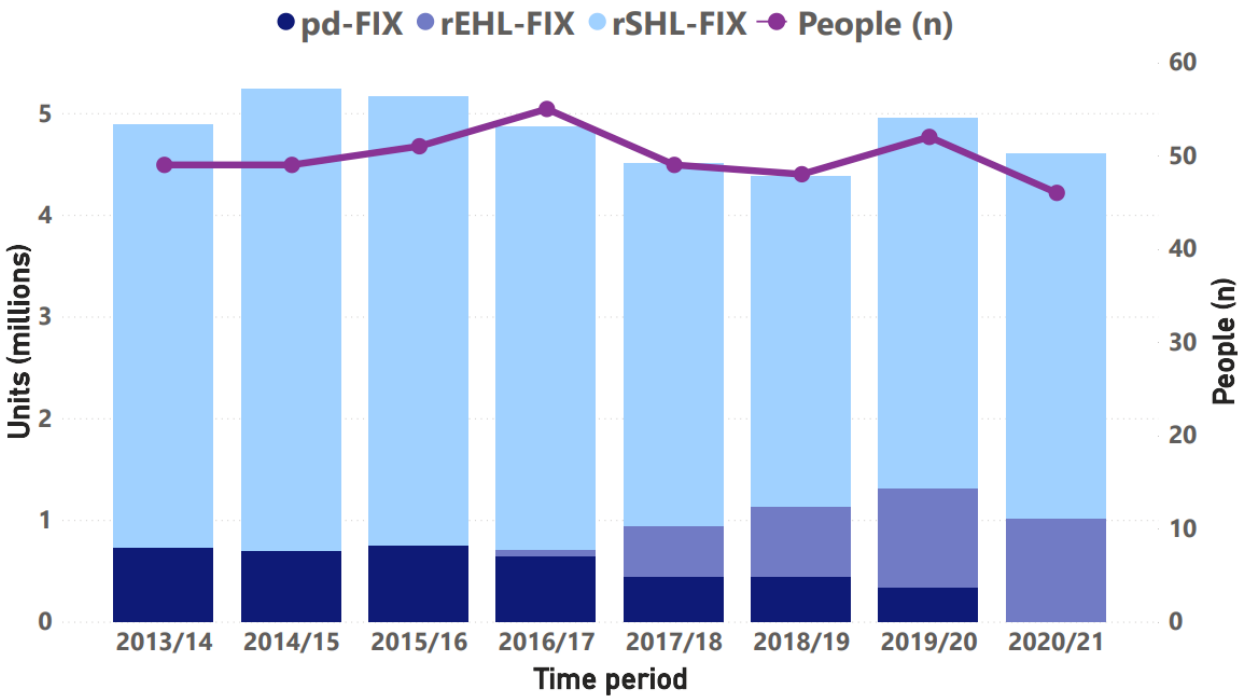


Figure 7c Factor IX units issued between April 2013 & March 2021 – *Severe Haemophilia B only*, people with a Scotland West postcode



Figures 7a to 7c give a historical view of the number of factor IX units issued between 2013/14 and 2020/21 for people with *severe haemophilia B only*. Figure 7a includes all people with a Scottish postcode. Figures 7b includes only people with a Scotland East postcode and Figure 7c those with a Scotland West postcode. The number of people treated is represented by the green line using a secondary axis.

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

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

Table 21a Data for Figure 7a - Factor IX units issued between April 2013 & March 2021 - *Severe Haemophilia B*, all Scottish postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	728,000	-	3,696,792	-	-	-	4,424,792	-	23	-
2014/2015	699,000	-3.98	3,705,000	+0.2	-	-	4,404,000	-0.47	25	+8.7
2015/2016	748,500	+7.1	4,434,250	+19.7	-	-	5,182,750	+17.7	24	-4.00
2016/2017	646,500	-13.63	3,293,000	-25.74	293,750	-	4,233,250	-18.32	24	-
2017/2018	442,500	-31.55	1,690,000	-48.68	1,011,250	+6.3	3,143,750	-25.74	24	-
2018/2019	441,000	-0.34	1,177,750	-30.31	1,230,250	+21.7	2,849,000	-9.38	23	-4.17
2019/2020	340,000	-22.90	1,252,250	+6.3	1,466,000	+19.2	3,058,250	+7.3	24	+4.3
2020/2021	-	-	1,270,000	+1.4	1,605,000	+9.5	2,875,000	-5.99	22	-8.33

Table 21b Data for Figure 7b - Factor IX units issued between April 2013 & March 2021 - *Severe Haemophilia B*, Scotland East postcodes

Year	Recombinant				Total		People treated	
	Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	2,375,542	-	-	-	2,375,542	-	10	-
2014/2015	1,953,000	-17.79	-	-	1,953,000	-17.79	11	+10
2015/2016	2,502,000	+28.1	-	-	2,502,000	+28.1	9	-18.18
2016/2017	1,536,000	-38.61	230,500	-	1,766,500	-29.40	9	-
2017/2018	360,500	-76.53	585,750	+154.1	946,250	-46.43	10	+11.1
2018/2019	66,000	-81.69	716,500	+22.3	782,500	-17.31	10	-
2019/2020	6,000	-90.91	746,500	+4.2	752,500	-3.83	10	-
2020/2021	-	-	737,250	-1.24	737,250	-2.03	9	-10.00

Table 21c Data for Figure 7c - Factor IX units issued between April 2013 & March 2021 - *Severe Haemophilia B*, Scotland West postcodes

Year	Plasma-derived		Recombinant				Total		People treated	
			Standard half-life		Enhanced half-life					
	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	IU	% difference year on year	n	% difference year on year
2013/2014	728,000	-	1,321,250	-	-	-	2,049,250	-	13	-
2014/2015	699,000	-3.98	1,752,000	+32.6	-	-	2,451,000	+19.6	15	+15.4
2015/2016	748,500	+7.1	1,932,250	+10.3	-	-	2,680,750	+9.4	15	-
2016/2017	646,500	-13.63	1,757,000	-9.07	63,250	-	2,466,750	-7.98	15	-
2017/2018	442,500	-31.55	1,329,500	-24.33	425,500	+572.7	2,197,500	-10.92	15	-
2018/2019	441,000	-0.34	1,111,750	-16.38	513,750	+20.7	2,066,500	-5.96	14	-6.67
2019/2020	340,000	-22.90	1,246,250	+12.1	719,500	+40	2,305,750	+11.6	15	+7.1
2020/2021	-	-	1,270,000	+1.9	867,750	+20.6	2,137,750	-7.29	13	-13.33

Von Willebrand Disease, Rarer Bleeding Disorders and Acquired Defects

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

Table 22 Concentrates issued to treat von Willebrand Disease

Manufacturer	Product	Patients Treated (n)	Total Units
CSL Behring	Voncento	67	811,000
Novo Nordisk	NovoSeven (mg)	1 - 2	176
	Desmopressin	53	1,257.7

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

Table 23 Concentrates issued to treat Rarer Bleeding Disorders

Manufacturer	Product	F.VII deficiency	F.IX deficiency	F.XIII deficiency
BPL	FXI	-	950	-
CSL Behring	Fibrogammin P	-	-	44,750
Novo Nordisk	NovoSeven (mg)	7	-	-
	NovoThirteen	-	-	62,500

Units in IU unless otherwise stated

Table 24 Concentrates issued to treat Acquired Defects

Manufacturer	Product	People treated (n)	Acquired haemophilia A (IU)	Acquired von Willebrands (IU)
CSL Behring	Voncento	3	-	48,500
Novo Nordisk	NovoSeven	1	279	-
Shire	OBIZUR	1	25,000	-
Takeda	Advate	1	46,500	-
	FEIBA	6	157,000	-

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

Table 25 Inhibitors by disease severity

Diagnosis	Severity (IU/dl) / subtype	In register *	Inhibitors		
			New n (%)	ongoing n (%)	Historical n (%)
Haemophilia A	< 1	165	0 (0.0)	13 (7.9)	18 (10.9)
	1 - 5	66	0 (0.0)	2 (3.0)	2 (3.0)
	> 5	290	0 (0.0)	1 (0.3)	4 (1.4)
	Total	**521**	**0 (0.0)**	**16 (3.1)**	**24 (4.6)**
Haemophilia B	< 1	26	0 (0.0)	1 (3.8)	0 (0.0)
	1 - 5	43	0 (0.0)	0 (0.0)	0 (0.0)
	> 5	67	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**136**	**0 (0.0)**	**1 (0.7)**	**0 (0.0)**
von Willebrand disease	Type 3	12	0 (0.0)	1 (8.3)	0 (0.0)
	Others	1,140	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**1,152**	**0 (0.0)**	**1 (0.1)**	**0 (0.0)**

** Including patients not regularly treated*

Table 25 shows the incidence of new inhibitors during 2020/21, 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 2020/21. Those labelled “ongoing” are those reported in previous years which have not been eradicated. Those reported as “historical” are those reported to have been previously eradicated or disappeared and not ongoing.

Table 26 Products issued to people with congenital bleeding disorders reported to have a positive inhibitor during 2020/21

Manufacturer	Product	People treated (n)	Units
Haemophilia A			
Alnylam Pharmaceuticals	ALN-AT3SC (Trial)	1	480
Baxter	Advate	1	300,000
	FEIBA	1	50,000
Novo Nordisk	Esperoct	1	10,000
	NovoEight	2	325,000
	NovoSeven (mg)	10	488
Pfizer	ReFacto AF	1	204,000
Roche	Hemlibra (mg)	11	44,190
Haemophilia B			
Alnylam Pharmaceuticals	ALN-AT3SC (Trial)	1	480
Novo Nordisk	NovoSeven (mg)	1	32
von Willebrand disease			
Novo Nordisk	NovoSeven (mg)	1	206
Co-inherited diagnoses			
Roche	Hemlibra (mg)	1	4,680

Units in IU unless otherwise stated

Table 26 shows the number of people with Scottish postcodes and reported products issued to those with an inhibitor, newly reported or ongoing during 2020/21, broken down by diagnosis and supplier.

Table 27 Adverse Events

Adverse event	Number of people	Number of events
Allergy event	1	1
Infection event	0	0
Inhibitor event	0	0
ICH event	4	4
Malignancy event	6	6
Neurological event	0	0
Other event	0	0
Poor efficacy event	0	0
Thrombotic event	3	3
Death event	16	16
COVID-19 event	5	5
Total	**35**	**35**

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

Table 27 shows the type and number of adverse events reported by Scottish centres during 2020/21.

Table 28 Causes of Death

Diagnosis	Cause of death	Severity by factor level (IU/dl)		
		1 - 5	> 5	Total
Haemophilia A	Carcinoma	0	2	**2**
	Unknown	0	2	**2**
Haemophilia B	Cerebral haemorrhage	0	1	**1**
	Ischaemic heart disease	1	0	**1**
	Unknown	0	1	**1**
Haemophilia A carrier	Infection (bacterial)	0	1	**1**
Acquired haemophilia A	Infection (bacterial)			**1**
	Stroke (unknown)			**1**
von Willebrand disease	Cerebral haemorrhage			**1**
	Ruptured aorta (peripheral vascular disease)			**1**
Acquired von Willebrands disease	Dementia/Alzheimer's disease			**1**
	Infection (bacterial)			**1**
Dysfibrinogenemia	Unknown			**1**
Other platelet defects	Carcinoma			**1**
	Total	**1**	**7**	**16**

Table 28 shows the causes of death reported in people with a Scottish postcode during 2020/21, broken down by diagnosis and disease severity.