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

Bleeding Disorder Statistics for Wales

April 2017 to March 2018

A report from the UK National Haemophilia Database

October 2018

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

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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	12	0	12
Haemophilia A Carrier		8	8
Acquired Haemophilia A	5	1	6
Haemophilia B	0	1	1
von Willebrand disease	6	15	21
Probable von Willebrands disease	1	1	2
Platelet-type Pseudo von Willebrand Disease	0	1	1
F.VII deficiency	0	3	3
F.X deficiency	0	1	1
F.XI Deficiency	7	9	16
Hypodysfibrinogenemia	1	0	1
Glanzmanns Thrombasthenia	1	0	1
Platelet defect	11	23	34
Unclassified	0	12	12
Total	**44**	**75**	**119**

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 Welsh 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	2	1	3	6
	10 : 19	1	0	0	1
	20 : 29	0	0	2	2
	30 : 39	0	0	0	0
	40 : 49	0	0	0	0
	50 : 59	0	0	1	1
	60 : 69	0	0	1	1
	70 +	0	0	1	1
	Total	**3**	**1**	**8**	**12**
Haemophilia B	0 : 9	0	0	0	0
	10 : 19	0	0	0	0
	20 : 29	0	0	0	0
	30 : 39	0	0	0	0
	40 : 49	0	0	0	0
	50 : 59	0	0	1	1
	60 : 69	0	0	0	0
	70 +	0	0	0	0
	Total	**0**	**0**	**1**	**1**

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 Welsh 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	279	1	280	139	49.64%
Acquired Haemophilia A	14	7	21	2	9.52%
Haemophilia A Carrier		66	66	4	6.06%
Haemophilia B	56	1	57	33	57.89%
Haemophilia B Carrier		16	16	1	6.25%
von Willebrand disease	155	276	431	39	9.05%
Acquired von Willebrands	5	0	5	2	40.00%
Probable von Willebrands disease	3	3	6	1	16.67%
Platelet-type Pseudo von Willebrand Disease	3	2	5	0	0.00%
Prothrombin Deficiency	0	1	1	0	0.00%
F.V deficiency	1	3	4	0	0.00%
F.VII deficiency	16	19	35	0	0.00%
F.X deficiency	0	2	2	0	0.00%
F.XI Deficiency	44	62	106	2	1.89%
F.XIII Deficiency	0	1	1	1	100.00%
Combined II+VII+IX+X Deficiency	1	0	1	0	0.00%
Co-inherited diagnoses	1	10	11	0	0.00%
Acquired Prothrombin Deficiency	1	0	1	0	0.00%
Afibrinogenemia	1	0	1	1	100.00%
Dysfibrinogenemia	8	16	24	0	0.00%
Hypofibrinogenemia	4	5	9	1	11.11%
Hypodysfibrinogenemia	1	1	2	0	0.00%
Glanzmanns Thrombasthenia	2	2	4	0	0.00%
Bernard Soulier	0	2	2	0	0.00%
Platelet Defect	47	92	139	1	0.72%
Miscellaneous	3	4	7	0	0.00%
Unclassified	4	28	32	0	0.00%
Totals	**649**	**620**	**1,269**	**227**	

Table 3 shows the total number of active registrations of patients with a Welsh 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 Health Board

Coagulation Defect	Abertawe Bro Morgannwg University	Aneurin Bevan	Betsi Cadwaladr University	Cardiff and Vale University	Cwm Taf	Hywel Dda	Powys Teaching	Total
Haemophilia A	63	61	51	49	26	25	5	280
Acquired Haemophilia A	6	1	5	4	1	4	0	21
Haemophilia A Carrier	26	8	15	9	3	5	0	66
Haemophilia B	6	17	6	12	11	4	1	57
Haemophilia B Carrier	3	4	1	4	2	1	1	16
von Willebrand disease	30	75	150	74	19	31	52	431
Acquired von Willebrands	0	3	0	2	0	0	0	5
Probable von Willebrands disease	0	2	1	2	0	1	0	6
Platelet-type Pseudo von Willebrand Disease	0	0	0	0	0	2	3	5
Prothrombin Deficiency	1	0	0	0	0	0	0	1
F.V deficiency	1	2	0	0	0	1	0	4
F.VII deficiency	8	2	14	2	0	8	1	35
F.X deficiency	0	0	1	1	0	0	0	2
F.XI Deficiency	16	49	11	13	7	9	1	106
F.XIII Deficiency	0	0	1	0	0	0	0	1
Combined II+VII+IX+X Deficiency	0	0	0	1	0	0	0	1
Co-inherited diagnoses	3	2	2	2	0	0	2	11
Acquired Prothrombin Deficiency	0	0	0	1	0	0	0	1
Afibrinogenemia	0	0	0	0	1	0	0	1
Dysfibrinogenemia	5	2	2	15	0	0	0	24
Hypofibrinogenemia	2	2	1	2	1	1	0	9
Hypodysfibrinogenemia	1	0	0	1	0	0	0	2
Glanzmanns Thrombasthenia	0	0	0	2	0	2	0	4
Bernard Soulier	0	0	1	1	0	0	0	2
Platelet Defect	34	21	29	29	15	7	4	139
Miscellaneous	1	1	2	1	1	0	1	7
Unclassified	5	10	2	10	3	2	0	32
Total	**211**	**262**	**295**	**237**	**90**	**103**	**71**	**1,269**

Table 4 shows the number of patients registered by health board. Patients are allocated to a health board based on their home postcode. The source database used for mapping postcodes to health boards can be found at http://geoportal.statistics.gov.uk/datasets/675f07b52292428992709d0af98d86d9_0.csv.

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	25	10	22	**57**
	≥18 years	57	22	144	**223**
	Total	**82**	**32**	**166**	**280**
Haemophilia B	<18 years	1	7	7	**15**
	≥18 years	10	15	17	**42**
	Total	**11**	**22**	**24**	**57**

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

Table 5 shows a more detailed breakdown of active registrations of patients with a Welsh 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	0	0	4	0	0	0	4	0
F.VII deficiency	1	0	34	0	0	0	35	0
F.X deficiency	0	0	2	0	0	0	2	0
F.XI Deficiency	7	0	99	2	0	0	106	2
Total	**8**	**-**	**139**	**2**	**-**	**-**	**147**	**2**

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

Table 6 shows the number of patients with selected rarer bleeding disorders and a Welsh 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	3	11	5	1	**20**	16	34	16	0	**66**	**86**	**3**
Type 2A	2	0	0	0	**2**	8	0	2	0	**10**	**12**	**3**
Type 2B	1	0	0	0	**1**	0	2	0	0	**2**	**3**	**1**
Type 2M	0	0	0	0	**0**	1	2	0	0	**3**	**3**	**0**
Type 2N	0	0	0	0	**0**	0	0	0	0	**0**	**0**	**0**
Type 2 Unspecified	0	1	0	0	**1**	1	0	0	0	**1**	**2**	**0**
Type 3	3				**3**	2				**2**	**5**	**4**
Type Unreported	1	1	1	1	**4**	13	13	14	0	**40**	**44**	**6**
Low VWF	0	0	0	0	**0**	0	0	0	0	**0**	**0**	**0**
										Sub Total Males	**155**	**17**
Females												
Type 1	5	8	4	0	**17**	18	36	53	0	**107**	**124**	**7**
Type 2A	0	1	0	0	**1**	11	6	5	0	**22**	**23**	**4**
Type 2B	0	0	0	0	**0**	0	1	5	0	**6**	**6**	**1**
Type 2M	0	2	1	0	**3**	6	1	3	0	**10**	**13**	**2**
Type 2N	0	0	0	0	**0**	0	0	4	0	**4**	**4**	**0**
Type 2 Unspecified	0	1	1	0	**2**	1	2	0	0	**3**	**5**	**1**
Type 3	0				**0**	2				**2**	**2**	**1**
Type Unreported	3	4	4	1	**12**	14	19	48	5	**86**	**98**	**6**
Low VWF	0	0	0	0	**0**	0	0	1	0	**1**	**1**	**0**
										Sub Total Females	**276**	**22**
										Grand Total - Males and Females	**431**	**39**

Table 7 shows patients registered with von Willebrand disease broken down by age, activity level, subtype, 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 Welsh cohort.

Treatment

Table 8 Patients with a Welsh 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	Birmingham & Black Country	1
	Bristol, North Somerset & South Gloucestershire	1
	Cheshire, Warrington & Wirral	18
	London	1
	Wales	121
	Sub total	**142**
Haemophilia A Carrier	Wales	4
	Sub total	**4**
Acquired Haemophilia A	Wales	2
	Sub total	**2**
Haemophilia B	Birmingham & Black Country	1
	Cheshire, Warrington & Wirral	3
	Wales	29
	Sub total	**33**
Haemophilia B Carrier	Wales	1
	Sub total	**1**
von Willebrand disease	Birmingham & Black Country	1
	Cheshire, Warrington & Wirral	2
	London	1
	Wales	36
	Sub total	**40**
Probable von Willebrand disease	Wales	1
Acquired von Willebrands	Wales	2
F.XI Deficiency	Wales	2
F.XIII Deficiency	Wales	1
Afibrinogenemia	Wales	1
Hypofibrinogenemia	Cheshire, Warrington & Wirral	1
Other platelet defects	Wales	1
	Grand total	**231**

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

Table 8 reports patients with a Welsh postcode by region which issued the treatment. Some patients received treatment outside of Wales. 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-Welsh postcode, registered & treated at a Welsh 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	East Midlands	1	1
	East of England	2	0
	London	2	2
	North East	0	1
	North West	1	0
	South East	3	2
	South West	7	3
	West Midlands	3	1
	Sub total	**19**	**10**
Haemophilia A Carrier	London	2	0
	South West	2	0
	West Midlands	1	0
	Sub total	**5**	**0**
Haemophilia B	London	1	0
	Yorkshire and the Humber	1	0
	Sub total	**2**	**0**
von Willebrand disease	East Midlands	3	1
	Greater Glasgow and Clyde	1	0
	South East	1	0
	South West	3	1
	West Midlands	3	0
	Yorkshire and the Humber	1	0
	Sub total	**12**	**2**
Acquired von Willebrands	London	0	1
F.XI Deficiency	South West	2	0
Platelet Defect	South West	1	0
Unclassified	South West	1	0
	Grand total	**42**	**13**

The patients reported in Table 9 were registered at or issued treatment from a Welsh Haemophilia Centre during 2017/18, however, they have a postcode recorded on the NHD which falls outside of Wales.

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	135	647,310	17,896,250	2,668,000	76,020	21,211,560
Haemophilia A Carrier	2	-	32,000	-	-	32,000
Acquired von Willebrands	2	75,000	3,000	-	-	78,000
von Willebrand disease	33	533,100	13,000	-	-	546,100
Total	**172**	**1,255,410**	**17,944,250**	**2,668,000**	**76,020**	**21,867,660**

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

Table 10 shows the number of patients with a Welsh 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	1,239,000
BPL	Optivate	394,810
CSL Behring	Helixate Nexgen	964,000
Grifols	Fanhdi	252,500
Novo Nordisk	NovoEight	627,500
	NovoSeven (mg)	690
Pfizer	ReFacto AF	8,805,250
Shire	Advate	6,260,500
	FEIBA	104,000
SOBI/Biogen	Elocta	2,668,000
	Investigational FVIII	76,020

Units in IU unless otherwise stated

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

Table 12 Factor VIII issued by Haemophilia Centre and Health Board for patients with *Severe* Haemophilia A (incl. treatment for inhibitors)

Haemophilia Centre Issuing Treatment	Health Board based on patients postcide	Severe Haemophilia A		
		Patients treated (n)	Total FVIII Units	Mean Usage
Bangor	Betsi Cadwaladr University Local Health Board	10	4,068,310	406,831
Birmingham (Queen Elizabeth)	Abertawe Bro Morgannwg University Local Health Board	1	231,000	231,000
Cardiff	Abertawe Bro Morgannwg University Local Health Board	13	2,772,770	213,290
	Aneurin Bevan Local Health Board	13	3,326,250	255,865
	Cardiff and Vale University Local Health Board	8	1,414,000	176,750
	Cwm Taf Local Health Board	7	1,148,750	164,107
	Hywel Dda Local Health Board	5	802,750	160,550
	Powys Teaching Local Health Board	1	354,000	354,000
Liverpool (R. I.)	Betsi Cadwaladr University Local Health Board	5	1,577,000	315,400
Liverpool Children's	Betsi Cadwaladr University Local Health Board	7	1,435,250	205,036
Manchester Children's	Betsi Cadwaladr University Local Health Board	1	5,000	5,000
Swansea	Abertawe Bro Morgannwg University Local Health Board	6	795,500	132,583
	Hywel Dda Local Health Board	2	490,500	245,250
	Totals	**79**	**18,421,080**	**233,178**

Table 12 reports the number of patients with a Welsh postcode 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 and by health board based on the patient's postcode as recorded on the NHD.

Note: Four patients were issued treatment from two different centres.

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
Powys Teaching Local Health Board	132,515	1	354,000	354,000	2.67
Betsi Cadwaladr University Local Health Board	696,284	21	7,085,560	337,408	10.18
Aneurin Bevan Local Health Board	587,743	13	3,326,250	255,865	5.66
Abertawe Bro Morgannwg University Local Health Board	531,858	18	3,799,270	211,071	7.14
Hywel Dda Local Health Board	384,239	7	1,293,250	184,750	3.37
Cardiff and Vale University Local Health Board	493,446	8	1,414,000	176,750	2.87
Cwm Taf Local Health Board	299,080	7	1,148,750	164,107	3.84
Wales	**3,125,165**	**75**	**18,421,080**	**1,683,951**	**5.89**

Ranked by mean usage

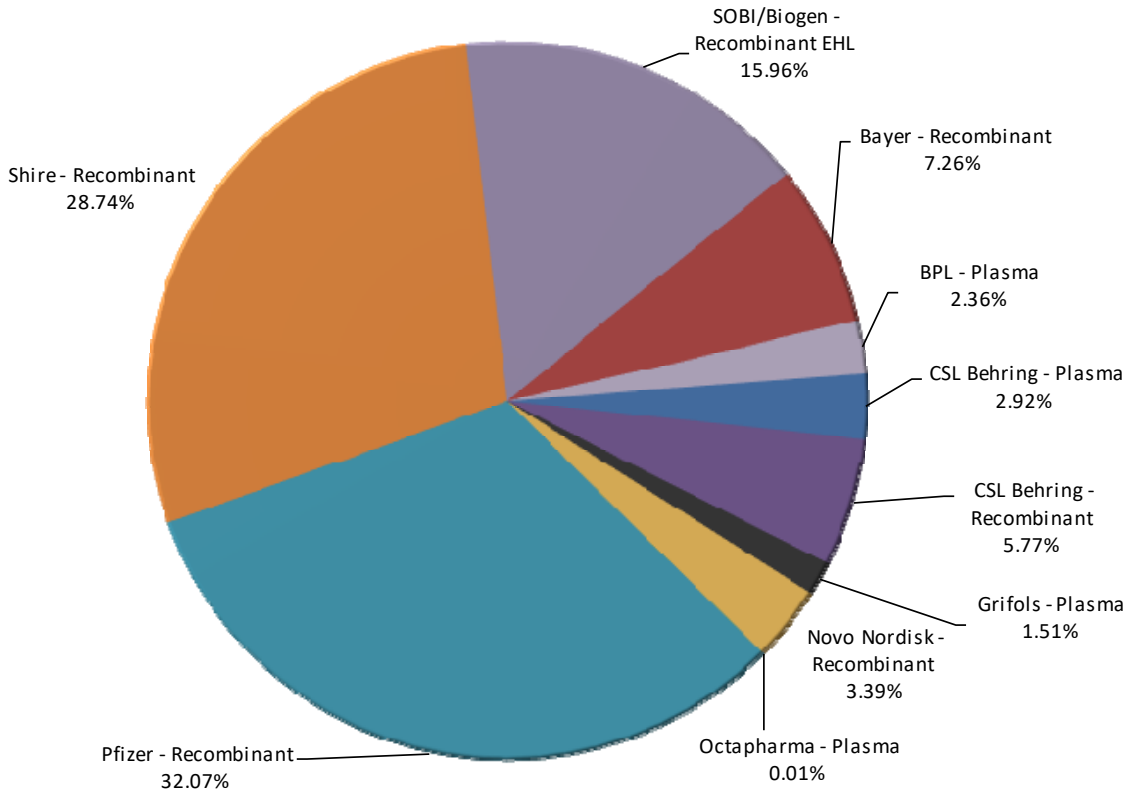
Mid-2017 population estimates, Office for National Statistics. Published by the Welsh Government, under the Open Government Licence v3.0. Updated June 2018

https://statswales.gov.wales/Catalogue/Population-and-Migration/Population/Estimates/Local-Health-Boards/populationestimates-by-lhb-age

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



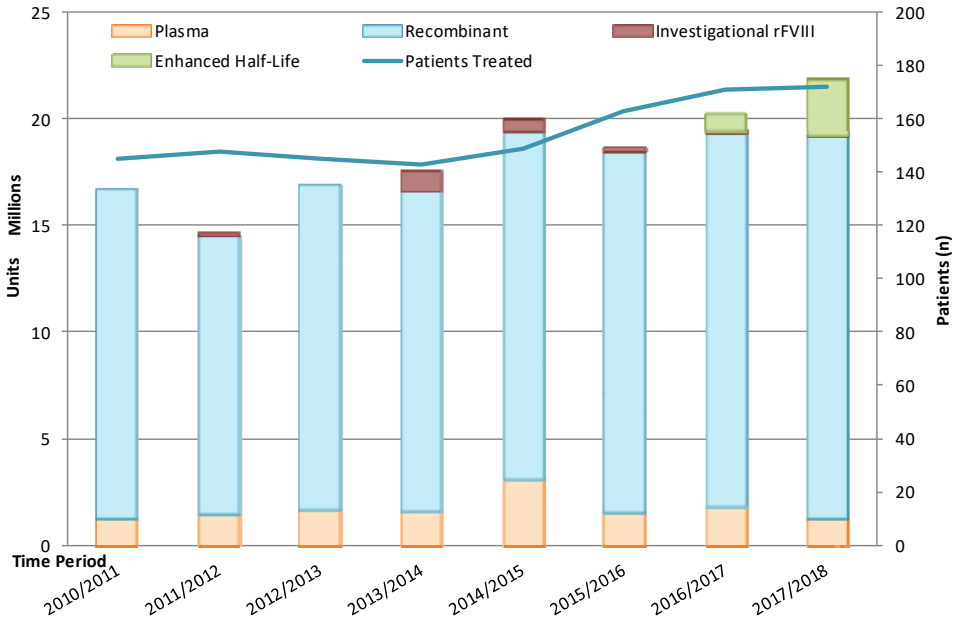
Includes products containing a combination of VWF and FVIII, which are reported in FVIII units

Manuacturer-Product Type	Patients (n)
Bayer - Recombinant	5
BPL - Plasma	1
CSL Behring - Plasma	32
CSL Behring - Recombinant	2
Grifols - Plasma	1
Novo Nordisk - Recombinant	13
Octapharma - Plasma	1
Pfizer - Recombinant	74
Shire - Recombinant	54
SOBI/Biogen - Recombinant EHL	14
Investigational FVIII	1
Total	**197**

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.

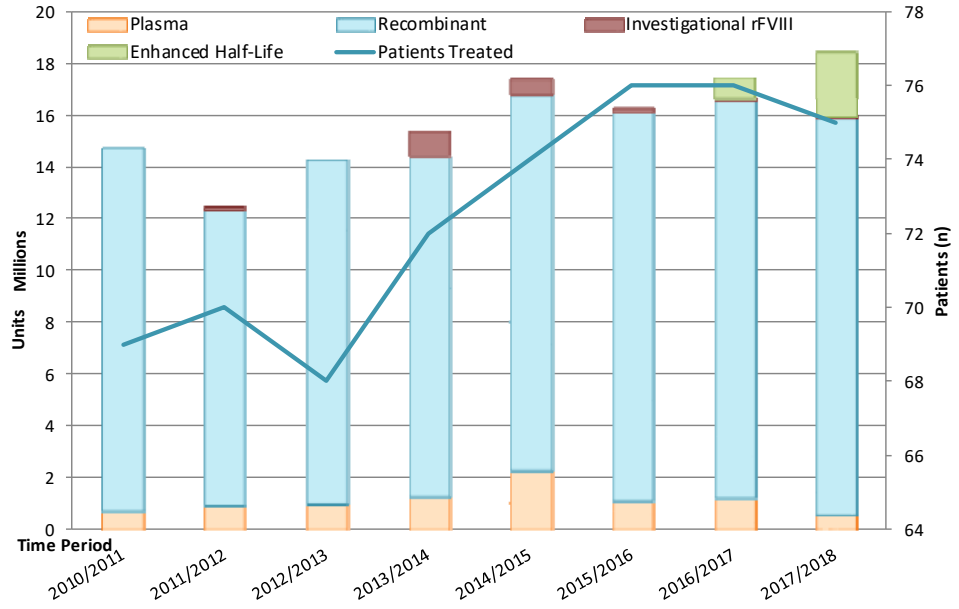
NOTE: The number of patients in this table cannot be compared with table 10 as this table includes patients treated with more than one product type. The patient numbers in Table 10 do not contain duplicates.

Figure 2 Factor VIII units issued by financial year between April 2010 & March 2018 – all diagnoses, all severities



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

Figure 3 Factor VIII units by financial year between April 2010 & March 2018 – *Severe Haemophilia A only*



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

Figures 2 and 3 give an historical view of the number of factor VIII units issued between 2010/11 and 2017/18 for all diagnoses and for patients with severe haemophilia A respectively. The number of patients treated is represented by the blue line using a secondary axis.

Table 14 Data table for Figure 2 - Factor VIII units issued by financial year between April 2010 & March 2018 - all diagnoses

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		IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	1,232,665	1.00	15,505,282	1.00	0		0		16,737,947	1.00	145	1.00
2011/2012	1,487,895	1.21	13,054,238	0.84	121,000	0.00	0		14,663,133	0.88	148	1.02
2012/2013	1,672,650	1.36	15,224,592	0.98	0	1.00	0		16,897,242	1.01	145	1.00
2013/2014	1,622,820	1.32	15,028,750	0.97	964,000	0.00	0		17,615,570	1.05	143	0.99
2014/2015	3,062,500	2.48	16,346,250	1.05	604,170	7.97	0		20,012,920	1.20	149	1.03
2015/2016	1,543,000	1.25	16,951,000	1.09	176,000	4.99	0		18,670,000	1.12	163	1.12
2016/2017	1,794,475	1.46	17,542,148	1.13	134,875	1.45	794,500		20,265,998	1.21	171	1.18
2017/2018	1,255,410	1.02	17,944,250	1.16	76,020	1.11	2,668,000		21,943,680	1.31	172	1.19

Table 15 Data table for Figure 3 - Factor VIII units by financial year between April 2010 & March 2018 - *Severe Haemophilia A*

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		IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	744,560	1.00	13,965,574	1.00	0		0		14,710,134	1.00	69	1.00
2011/2012	930,000	1.25	11,447,738	0.82	121,000	1.00	0		12,498,738	0.85	70	1.01
2012/2013	1,009,150	1.36	13,238,592	0.95	0	0.00	0		14,247,742	0.97	68	0.99
2013/2014	1,275,000	1.71	13,129,000	0.94	964,000	7.97	0		15,368,000	1.04	72	1.04
2014/2015	2,291,500	3.08	14,453,000	1.03	604,170	4.99	0		17,348,670	1.18	74	1.07
2015/2016	1,135,000	1.52	14,946,250	1.07	176,000	1.45	0		16,257,250	1.11	76	1.10
2016/2017	1,236,475	1.66	15,295,000	1.10	134,875	1.11	769,500		17,435,850	1.19	76	1.10
2017/2018	647,310	0.87	15,242,750	1.09	76,020	0.63	2,455,000		18,421,080	1.25	75	1.09

Tables 14 and 15 are the data tables for Figures 2 and 3.

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	33	78,000	1,824,500	678,500	-	2,581,000
Haemophilia B Carrier	1	-	42,000	-	-	42,000
Total	**34**	**78,000**	**1,866,500**	**678,500**	**-**	**2,623,000**

Table 16 shows the number of patients with a Welsh 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
CSL Behring	IDELVION	58,000
	Mononine	78,000
Pfizer	Benefix	1,733,500
Shire	RIXUBIS	91,000
SOBI/Biogen	ALPROLIX	620,500

Units in IU unless otherwise stated

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

Table 18 Factor IX issued to patients with *Severe* Haemophilia B (incl. treatment for inhibitors), by Haemophilia Centre and Health Board

Haemophilia Centre Issuing Treatment	Health Board based on patients postcide	Severe Haemophilia B		
		Patients treated (n)	Total FIX Units	Mean Usage
Cardiff	Abertawe Bro Morgannwg University Local Health Board	1	114,000	114,000
	Aneurin Bevan Local Health Board	3	400,000	133,333
	Cwm Taf Local Health Board	4	590,500	147,625
Swansea	Abertawe Bro Morgannwg University Local Health Board	1	192,000	192,000
	Totals	**9**	**1,296,500**	**144,056**

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 and by health board based on the patient's postcode as recorded on the NHD.

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

Health Board	General Population	Severe Haemophilia B			
		Patients treated (n)	Total FIX Units (IU)	Mean Usage	FIX Units Per Capita
Abertawe Bro Morgannwg University Local Health Board	531,858	2	306,000	153,000	0.58
Cwm Taf Local Health Board	299,080	4	590,500	147,625	1.97
Aneurin Bevan Local Health Board	587,743	3	400,000	133,333	0.68
Betsi Cadwaladr University Local Health Board	696,284	0	0	-	0.00
Cardiff and Vale University Local Health Board	493,446	0	0	-	0.00
Hywel Dda Local Health Board	384,239	0	0	-	0.00
Powys Teaching Local Health Board	132,515	0	0	-	0.00
Wales	**3,125,165**	**9**	**1,296,500**	**433,958**	**3.23**

Ranked by mean usage

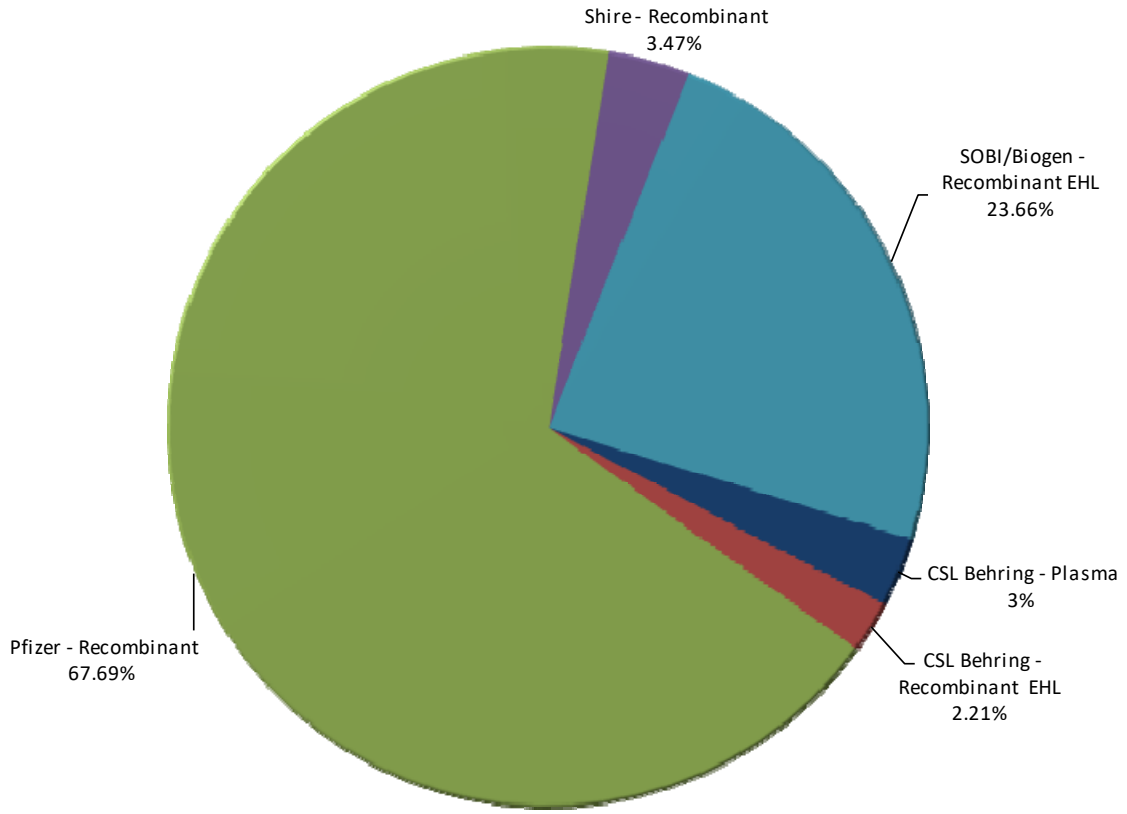
Mid-2017 population estimates, Office for National Statistics. Published by the Welsh Government, under the Open Government Licence v3.0. Updated June 2018

https://statswales.gov.wales/Catalogue/Population-and-Migration/Population/Estimates/Local-Health-Boards/populationestimates-by-lhb-age

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 Welsh postcode between April 2017 & March 2018



Manuacturer-Product Type	Patients (n)
CSL Behring - Plasma	2
CSL Behring - Recombinant EHL	2
Pfizer - Recombinant	28
Shire - Recombinant	2
SOBI/Biogen - Recombinant EHL	5
Total	**39**

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.

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 5 Factor IX units by financial year between April 2010 & March 2018 - all diagnoses, all severities

Figure 5 gives an historical view of the number of factor IX units issued between 2010/11 and 2017/18 for all diagnoses. The number of patients treated is represented by the blue line using a secondary axis.

Table 20 Data table for figure 5 - Factor IX units issued by financial year between April 2010 & March 2018 – all diagnoses

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		IU	% difference since 2010/11	n	% difference since 2010/11
2010/2011	212,000	1.00	2,077,000	1.00	0		0		2,289,000	1.00	27	1.00
2011/2012	106,000	0.50	2,203,500	1.06	647,689	1.00	0		2,957,189	1.29	28	1.04
2012/2013	133,000	0.63	1,915,500	0.92	517,180	0.80	0		2,565,680	1.12	25	0.93
2013/2014	274,000	1.29	2,326,770	1.12	349,016	0.54	0		2,949,786	1.29	34	1.26
2014/2015	198,000	0.93	2,212,000	1.06	617,447	0.95	0		3,027,447	1.32	27	1.00
2015/2016	228,000	1.08	2,281,000	1.10	136,553	0.21	0		2,645,553	1.16	30	1.11
2016/2017	170,000	0.80	2,188,000	1.05	263,692	0.41	289,015		2,910,707	1.27	30	1.11
2017/2018	78,000	0.37	1,866,500	0.90	0	0.00	678,500		2,623,000	1.15	34	1.26

Table 20 is the data table for Figure 5.

Von Willebrand Disease, Rarer Bleeding Disorders and Acquired Defects

Tables 21 - 23 shows the number of patients with Welsh 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
Bayer	Kogenate (n= 1)	13,000
CSL Behring	Voncento (n= 31)	531,100
LFB Biomedicaments	Willfact /Wilfactin (n= 1)	17,000
Octapharma	Wilate (n= 1)	2,000
Desmopressin	Desmopressin (n= 6)	153.6

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.XI Deficiency	F.XIII Deficiency
BPL	FXI (n= 1)	1,000	-
CSL Behring	Fibrogammin P (n= 1)	-	15,000
Octapharma	Octaplas (units) (n= 1)	4	-

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= 1)	-	75,000
Pfizer	ReFacto AF (n= 1)	-	3,000
Shire	FEIBA (n= 2)	128,000	-

Units in IU unless otherwise stated
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	82	0 (0.0)	10 (12.2)	15 (18.3)
	1 - 5	32	0 (0.0)	0 (0.0)	2 (6.3)
	> 5	166	0 (0.0)	2 (1.2)	4 (2.4)
	Total	**280**	**0 (0.0)**	**12 (4.3)**	**21 (7.5)**
Haemophilia B	< 1	11	0 (0.0)	0 (0.0)	0 (0.0)
	1 - 5	22	0 (0.0)	0 (0.0)	0 (0.0)
	> 5	24	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**57**	**0 (0.0)**	**0 (0.0)**	**0 (0.0)**
von Willebrand disease	Type 3	7	0 (0.0)	1 (14.3)	0 (0.0)
	Others	424	0 (0.0)	0 (0.0)	0 (0.0)
	Total	**431**	**0 (0.0)**	**1 (0.2)**	**0 (0.0)**

** 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		
Bayer	Kogenate (n= 1)	224,000
BPL	Optivate (n= 1)	394,810
CSL Behring	Helixate Nexgen (n= 1)	863,000
Grifols	Fanhdi (n= 1)	252,500
Novo Nordisk	NovoSeven (mg) (n= 4)	690
Pfizer	ReFacto AF (n= 1)	276,000
Shire	Advate (n= 3)	447,500
	FEIBA (n= 1)	104,000
SOBI/Biogen	Elocta (n= 4)	1,412,500
von Willebrand Disease		
CSL Behring	Voncento (n= 1)	78,800

Units in IU unless otherwise stated

Table 25 shows the number of patients with Welsh 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	1
Infection Event	0
Inhibitor Event	0
Intracranial haemorrhage	0
Malignancy Event	1
Other Event	1
Poor Efficacy Event	0
Thrombotic Event	0
Total	**3**

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 Welsh postcode during 2017/18.

Table 27 Causes of Death

Coagulation Defect	Cause of Death	Severity (factor level iu/dl)				
		< 1	1 - 5	> 5	N/K	Total
Haemophilia A	Unknown	0	1	0	**0**	**1**
Acquired Haemophilia A	Infection (Bacterial)	1	0	2	**1**	**4**
von Willebrand disease	Carcinoma	0	1	0	**0**	**1**
	Liver Failure	0	0	1	**0**	**1**
	Unknown	0	0	2	**0**	**2**
F.XI Deficiency	Unknown	0	0	1	**0**	**1**
Platelet Defect	Unknown					**3**
	Total	**1**	**2**	**6**	**1**	**13**

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