

# Bleeding Disorder Statistics for April 2015 to March 2016

A report from the UK National Haemophilia Database

# Haemophilia A

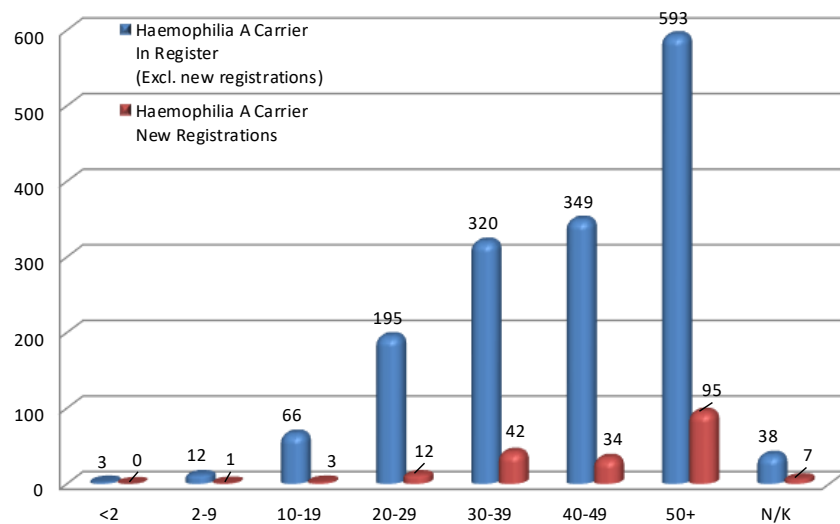
**Table 1 Patients with congenital Haemophilia A registered and treated April 2015 - March 2016**

Haemophilia A	Age Range	Factor VIII level (iu/dl)					
		< 1	1 - 5	>5 & <40	≥ 40	Unknown	Total
Total In Register (including low-level carriers)	<18 years	665 (0)	219 (4)	690 (87)	136 (113)	9 (8)	1,719 (212)
	≥18 years	1,182 (2)	669 (2)	2,983 (559)	990 (840)	157 (155)	5,981 (1558)
	<b>Total</b>	<b>1,847 (2)</b>	<b>888 (6)</b>	<b>3,673 (646)</b>	<b>1,126 (953)</b>	<b>166 (163)</b>	<b>7,700 (1770)</b>
New Registrations * (including low-level carriers)	<18 years	57 (0)	11 (0)	67 (15)	19 (18)	4 (3)	158 (36)
	≥18 years	16 (0)	4 (0)	108 (43)	100 (91)	25 (24)	253 (158)
	<b>Total</b>	<b>73 (0)</b>	<b>15 (0)</b>	<b>175 (58)</b>	<b>119 (109)</b>	<b>29 (27)</b>	<b>411 (194)</b>
Treated in year** (including low-level carriers)	<18 years	638 (0)	173 (3)	157 (7)	6 (2)	1 (0)	975 (12)
	≥18 years	1,134 (0)	464 (2)	663 (48)	38 (17)	2 (1)	2,301 (68)
	<b>Total</b>	<b>1,772 (0)</b>	<b>637 (5)</b>	<b>820 (55)</b>	<b>44 (19)</b>	<b>3 (1)</b>	<b>3,276 (80)</b>

\* New registrations are a subset of the 'In Register' numbers

\*\* Treated includes patients 'In Register' and 'New Registrations'

**Figure 1** Carriers of Haemophilia A currently registered and newly registered, by baseline factor VIII level



Diagnosis	Factor VIII level (iu/dl)								Grand Total
	<2	2-9	10-19	20-29	30-39	40-49	50+	N/K	
Haemophilia A Carrier In Register (Excl. new registrations)	3	12	66	195	320	349	593	38	1576
Haemophilia A Carrier New Registrations	0	1	3	12	42	34	95	7	194
<b>Total</b>	<b>3</b>	<b>13</b>	<b>69</b>	<b>207</b>	<b>362</b>	<b>383</b>	<b>688</b>	<b>45</b>	<b>1770</b>

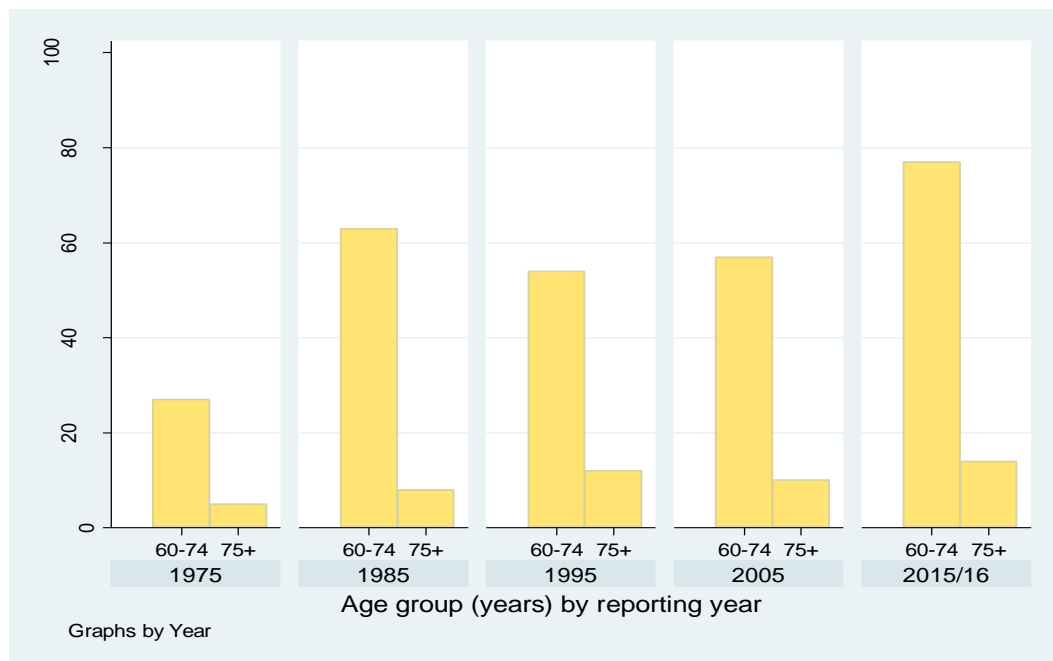
**Table 2**      **New registrations of Haemophilia A between April 2015 & March 2016, by age at mid-year and severity**

Coagulation Defect	Age (years)	Number of Patients (Factor VIII level (iu/dl))					
		< 1	1 - 5	>5 & <40	≥ 40	Unknown	Total
Haemophilia A	0 : 4	52	10	23	1	1	87
	5 : 9	3	1	13	0	0	17
	10 : 19	2	0	20	1	0	23
	20 : 29	13	2	16	2	1	34
	30 : 39	3	2	10	1	0	16
	40 : 49	0	0	9	2	0	11
	50 : 59	0	0	7	1	0	8
	60 : 69	0	0	8	1	0	9
	70 +	0	0	11	1	0	12
<b>Total</b>		<b>73</b>	<b>15</b>	<b>117</b>	<b>10</b>	<b>2</b>	<b>217</b>

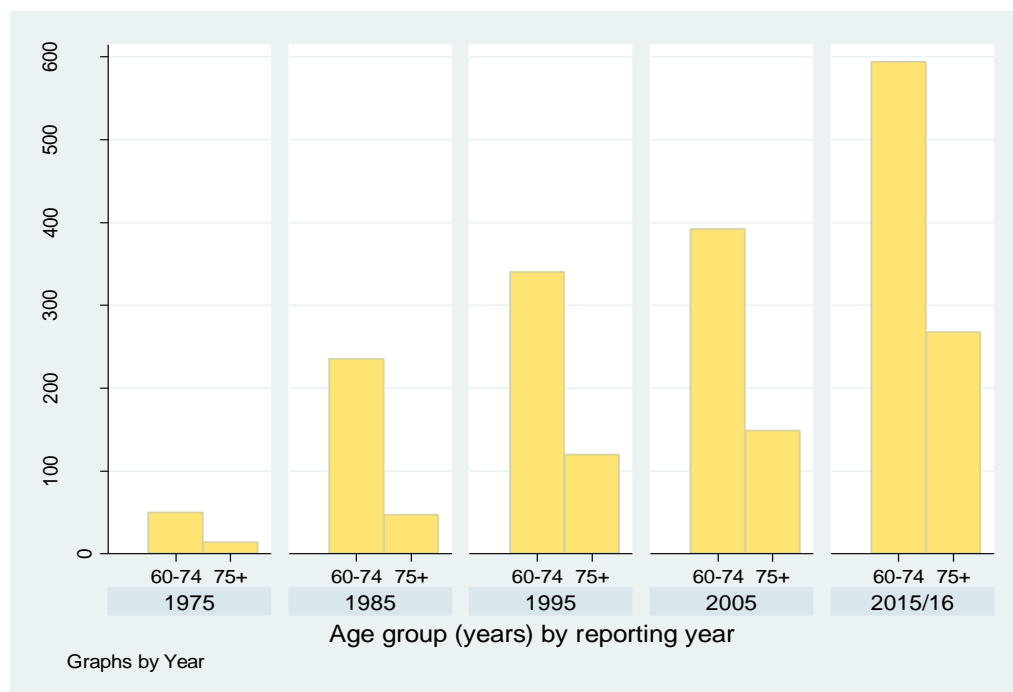
**Table 3** New registrations of patients with Severe Haemophilia A aged over 5 years old, and subsequent treatment by year

Haemophilia A	Registration year								Total 2008/2016
	2008/09	2009/10	2010/11	2011/12	2012/13	2013/14	2014/15	2015/16	
New registrations per year	12	10	11	15	11	17	19	23	118
Treated in each year									
2008/09	12								12
2009/10	11	6							17
2010/11	10	8	10						28
2011/12	9	8	9	14					41
2012/13	8	8	7	14	10				53
2013/14	8	8	8	12	9	16			63
2014/15	7	8	8	12	9	16	18		79
2015/16	7	8	6	10	8	15	19	20	93
No treatment record	0	1	1	0	1	0	0	3	6

**Figure 2** Trend in numbers of Severe Haemophilia A patients aged 60 years and above, 1975 - 2015/16 (including HIV +ve pts)

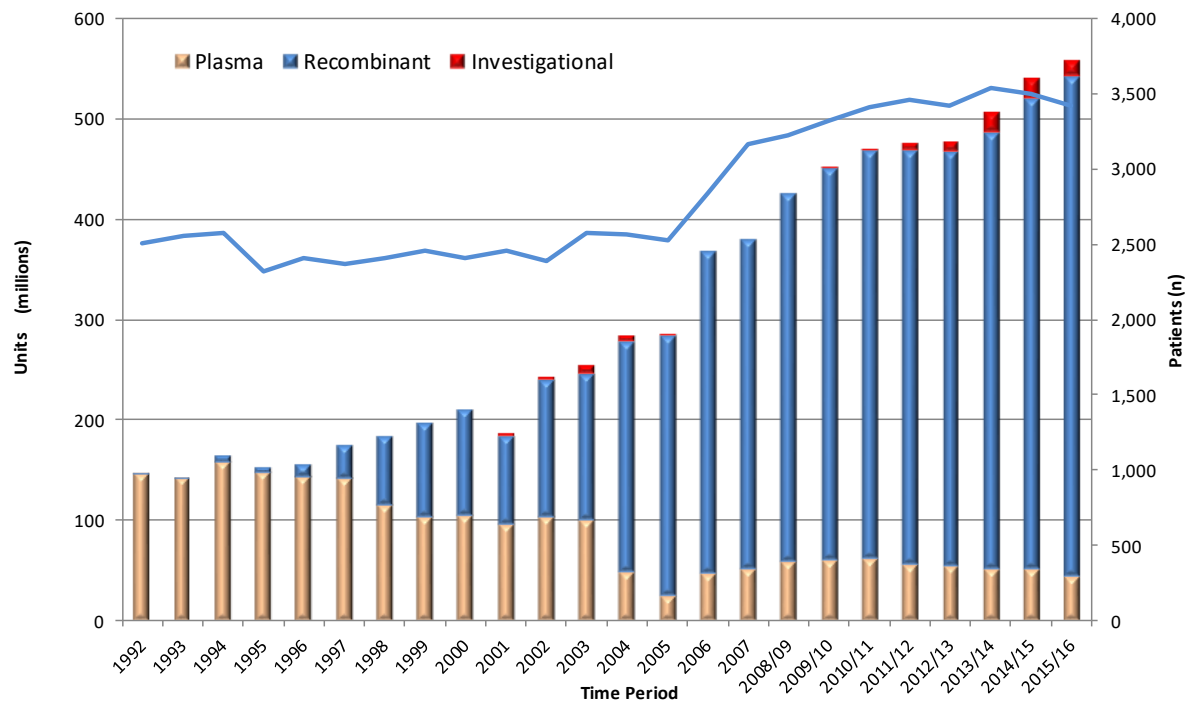


**Figure 3** Trend in numbers of non-Severe Haemophilia A patients aged 60 years and above, 1975 - 2015/16 (including HIV +ve pts)



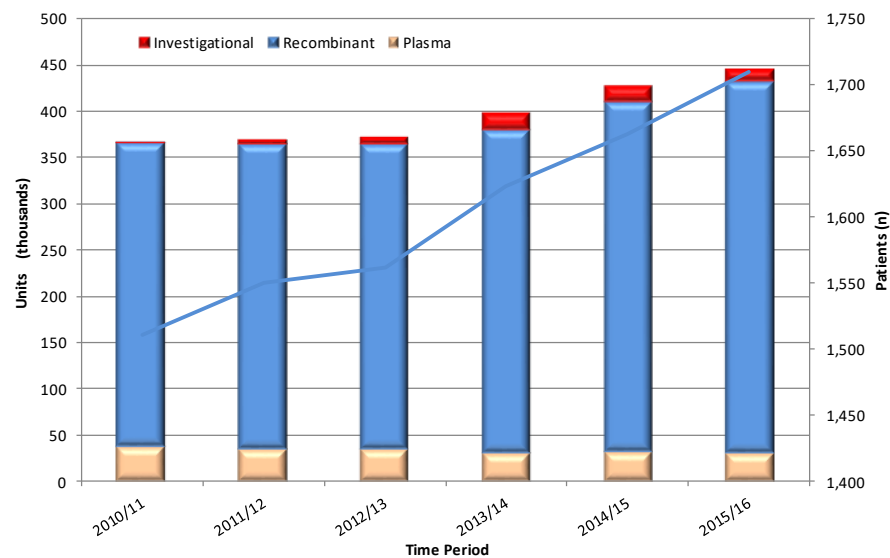


**Figure 4** Factor VIII units used by UK Haemophilia Centres 1992 - 2015/16 all diagnoses



*N.B: Data for St Thomas' were not submitted 1996-2006.*

**Figure 5** Factor VIII units used by UK Haemophilia Centres to treat Severe Haemophilia A - 2010/11 - 2015/16



Year	Plasma		Recombinant (excluding investigational)		Investigational rFVIII		Total		Patients	
	IU (thousands)	% difference since 2010/11	IU (thousands)	% difference since 2010/11	IU (thousands)	% difference since 2010/11	IU (thousands)	% difference since 2010/11	n	% difference since 2010/11
2010/11	37,185	100.00%	327,010	100.00%	1,123	100.00%	365,318	100.00%	1,511	100.00%
2011/12	33,846	91.02%	329,137	100.65%	6,645	591.49%	369,629	101.18%	1,550	102.58%
2012/13	33,358	89.71%	330,012	100.92%	8,116	722.50%	371,487	101.69%	1,562	103.38%
2013/14	30,189	81.19%	348,973	106.72%	18,484	1645.39%	397,646	108.85%	1,623	107.41%
2014/15	31,438	84.54%	377,822	115.54%	17,496	1557.45%	426,756	116.82%	1,663	110.06%
2015/16	30,094	80.93%	400,601	122.50%	14,198	1263.90%	444,893	121.78%	1,710	113.17%

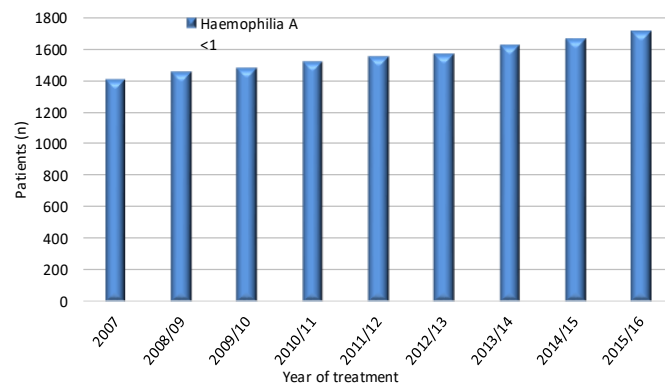
**Table 4 Treatment intensity of patients with Severe Haemophilia A - 2010/11 - 2015/16**

Treatment Period	FVIII Units		Patients (n)		Treatment Intensity (Units/Pt)		Change in treatment intensity since 2010/11 (%)	
	<18 Years	≥18 years	<18 Years	≥18 years	<18 Years	≥18 years	<18 Years	≥18 years
2010/11	131,141,658	234,176,810	607	904	216,049	259,045	100.0	100.0
2011/12	128,016,899	241,611,622	597	953	214,434	253,527	99.3	97.9
2012/13	125,289,759	246,197,158	605	957	207,091	257,259	95.9	99.3
2013/14	131,810,117	265,836,344	625	998	210,896	266,369	97.6	102.8
2014/15	140,999,503	285,756,508	622	1,041	226,687	274,502	104.9	106.0
2015/16	139,722,386	305,170,204	628	1,082	222,488	282,043	103.0	108.9

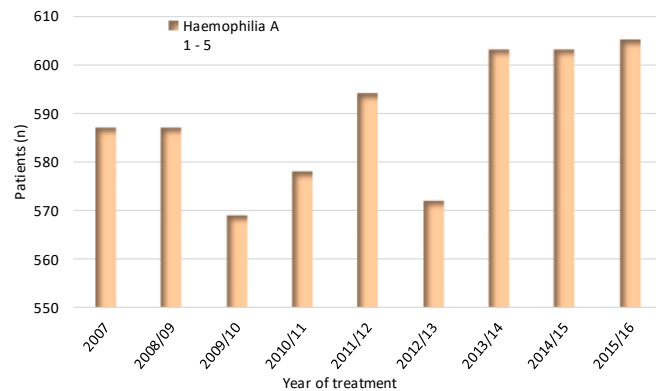
*Treatment includes Plasma, Recombinant, Investigational factor VIII and ITI usage*

**Figures 6a, b & c Patients with Severe / Moderate / Mild Haemophilia A treated with FVIII by UK Haemophilia Centres - 2007 - 2015/16**

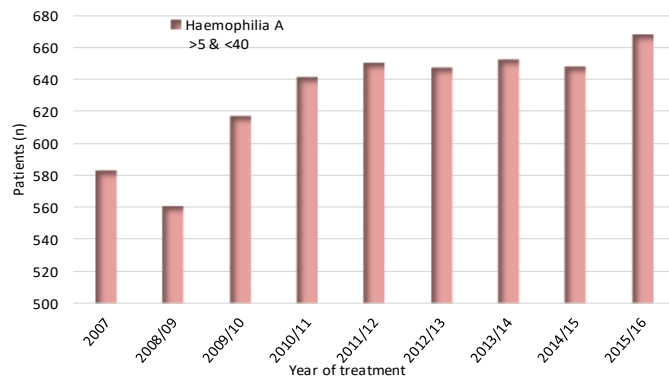
**Figure 6a**



**Figure 6b**



**Figure 6c**



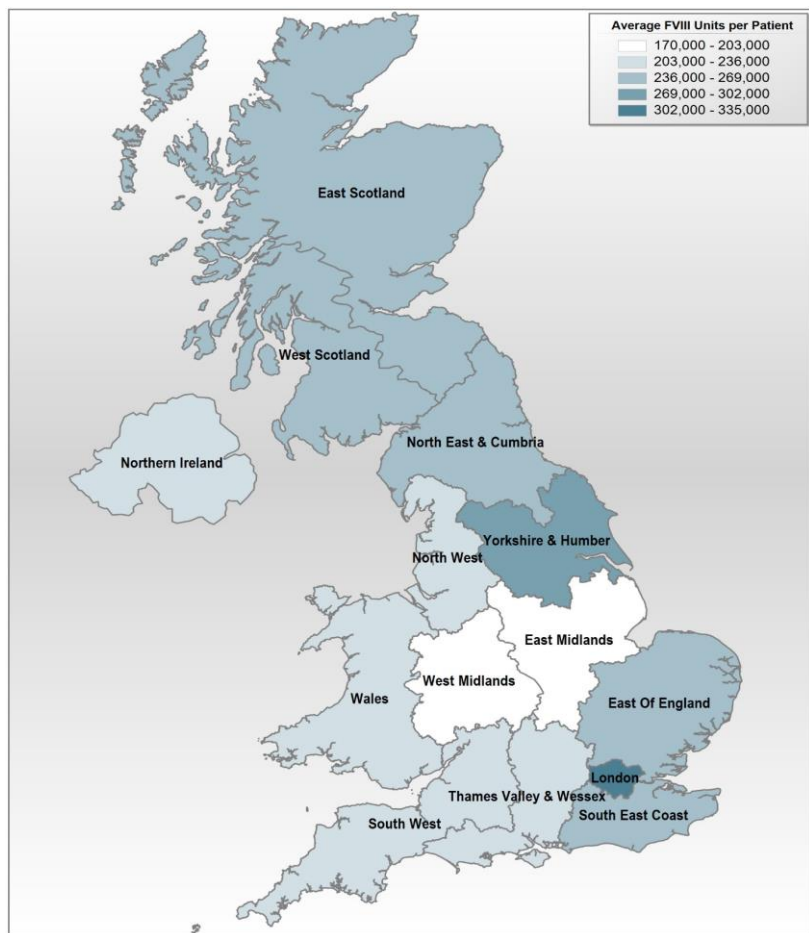
Data table for Figures 6a, 6b & 6c

Treatment Year	Haemophilia A <1		Haemophilia A 1 - 5		Haemophilia A >5 & <40		Haemophilia A ≥ 40	
	n	% difference since 2007	n	% difference since 2007	n	% difference since 2007	n	% difference since 2007
2007	1405	100.0	587	100.0	583	100.0	26	100.0
2008/09	1454	103.5	587	100.0	561	96.2	22	84.6
2009/10	1475	105.0	569	96.9	617	105.8	26	100.0
2010/11	1511	107.5	578	98.5	641	109.9	26	100.0
2011/12	1550	110.3	594	101.2	650	111.5	20	76.9
2012/13	1562	111.2	572	97.4	647	111.0	19	73.1
2013/14	1623	115.5	603	102.7	652	111.8	19	73.1
2014/15	1663	118.4	603	102.7	648	111.1	21	80.8
2015/16	1710	121.7	605	103.1	668	114.6	21	80.8

**Figure 7**      **Number of Severe Haemophilia A patients treated per million capita**



**Figure 8** Factor VIII usage (including Inhibitor) divided per Severe Haemophilia A patients by commissioning region



**Table 5 Factor VIII usage by region for patients with Severe Haemophilia A (incl. treatment for inhibitors)**

Region	General Population	Patients treated (n) Severe Haemophilia A	Total FVIII Units (IU) Severe Haemophilia A	Mean Usage Severe Haemophilia A	FVIII Units Per Capita Severe Haemophilia A
London		491	158,483,666	322,777	
South Yorkshire & Bassetlaw		118	35,383,750	299,862	
Cumbria, Northumberland & Tyne and Wear		63	16,678,500	264,738	
East Anglia		60	15,568,000	259,467	
Surrey & Sussex		34	8,434,500	248,074	
Wessex		232	53,075,594	228,774	
Cheshire, Warrington & Wirral		146	30,905,250	211,680	
Bristol, North Somerset & South Gloucestershire		77	16,087,750	208,932	
Leicestershire & Lincolnshire		91	16,300,727	179,129	
Birmingham & Black Country		146	25,330,000	173,493	
<b>England</b>	<b>54,786,300</b>	<b>1,423</b>	<b>376,247,737</b>	<b>264,405</b>	<b>6.87</b>
Northern Ireland	1,851,600	69	15,524,750	224,996	8.38
Scotland East	3,021,290	85	22,173,000	260,859	7.34
Scotland West	2,351,710	56	13,327,500	237,991	5.67
Wales	3,099,100	83	17,619,603	212,284	5.69
<b>United Kingdom</b>	<b>65,110,000</b>	<b>1,711</b>	<b>444,892,590</b>	<b>260,019</b>	<b>6.83</b>

English regions ranked by mean usage

**Source accreditation:**

England, Northern Ireland & Wales: Mid-2015 Population estimate: Office for National Statistics licensed under the Open Government Licence v.3.0.

Scotland: Mid-2015 Population Estimates supplied by National Records of Scotland under the Open Government Licence v3.0 © Crown Copyright 2016

The English commissioning area teams have been mapped to the following geographical regions:

Area Team Name	Geographical Region Name
Birmingham and the Black Country	West Midlands
Bristol, North Somerset and South Gloucestershire	South West
Cheshire, Warrington and Wirral	North West
Cumbria, Northumberland, Tyne and Wear	North East & Cumbria
East Anglia	East of England
Leicestershire and Lincolnshire	East Midlands
London	London
South Yorkshire and Bassetlaw	Yorkshire & Humber
Surrey and Sussex	South East Coast
Wessex	Thames Valley & Wessex

N.B:

Patients resident in England are reported against all English commissioning areas in which they were treated, so may be counted more than once.

The total number of severe Haemophilia A patients treated in the UK in 2015/16 was 1711.

Patients resident in England and treated in Wales, Scotland or Northern Ireland are reported against the devolved administration in which they were treated.

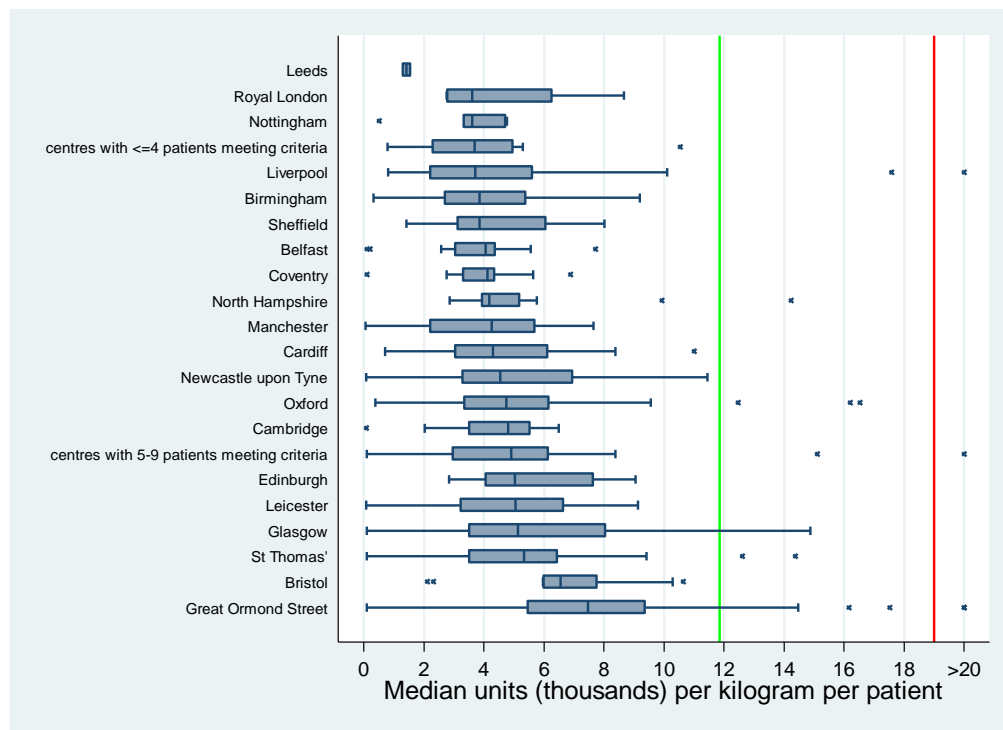
Patients resident in Wales, Scotland or Northern Ireland are reported against their devolved administrations, regardless of where they were treated.

Patients resident in Scotland are reported against East/West Scotland according to their home postcode, rather than where they were treated.

Patients resident outside the UK are reported against the commissioning areas in which they were treated.



**Figure 9** Annual FVIII usage 2015/16 in Severe Haemophilia A patients aged under 18 years with no current inhibitor, by centre, ranked by median usage



*N.B:*

*Patients treated at more than 1 centre during the year have all their usage reported against the centre where they received the most units*

*Indicated outside values show where a patient's usage is more than 1.5 times the interquartile range (IQR) from the nearest quartile at centre level*

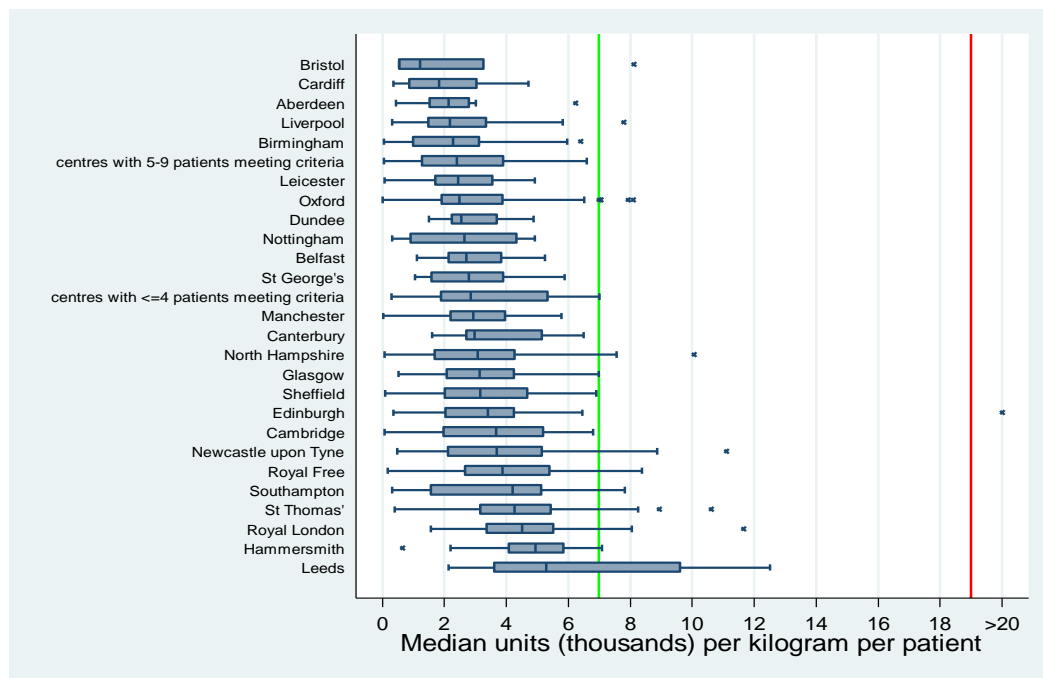
*The vertical green line indicates the 95<sup>th</sup> percentile of units, regardless of centre (11,852 IU/kg)*

*The vertical red line indicates a break in the axis in order to illustrate extreme outliers*

Data table for Figure 9

Haemophilia Centre	Patients treated	Patients treated with weight reported	Total Units	Median Units	Median Units/Kg
Belfast	18	18	2,827,750	155,375	4,049
Birmingham	39	28	5,244,750	130,000	3,844
Bristol	23	12	4,672,250	157,000	6,545
Cambridge	23	22	3,077,000	124,500	4,806
Cardiff	10	9	1,734,750	175,000	4,290
Coventry	10	10	1,679,750	140,750	4,112
Edinburgh	11	9	1,848,000	98,500	5,026
Glasgow	21	21	3,720,750	157,500	5,125
Great Ormond Street	77	76	19,965,240	245,000	7,462
Leeds	24	2	4,905,500	184,875	1,424
Leicester	10	8	958,200	96,375	5,056
Liverpool	22	22	2,984,750	100,500	3,708
Manchester	42	40	6,398,000	132,750	4,252
Newcastle upon Tyne	16	16	3,524,500	220,500	4,552
North Hampshire	19	19	3,102,250	165,000	4,171
Nottingham	19	5	1,999,500	87,500	3,609
Oxford	39	37	8,526,750	183,000	4,752
Royal London	13	7	2,234,500	168,000	3,607
Sheffield	15	10	2,855,250	111,500	3,856
St Thomas'	22	13	3,641,000	140,875	5,329
Centres with <=4 patients meeting criteria	16	11	2,955,250	167,500	3,683
Centres with 5-9 patients meeting criteria	61	56	13,617,100	190,000	4,911

**Figure 10** Annual FVIII usage 2015/16 in Severe Haemophilia A patients aged 18 years or more with no current inhibitor, by centre, ranked by median usage



*N.B:*

*Patients treated at more than 1 centre during the year have all their usage reported against the centre where they received the most units*

*Indicated outside values show where a patient's usage is more than 1.5 times the IQR from the nearest quartile at centre level*

*The vertical green line indicates the 95<sup>th</sup> percentile of units, regardless of centre (6,980 IU/kg)*

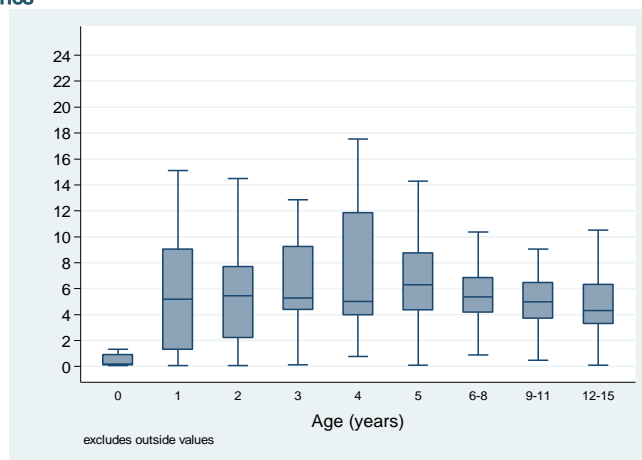
*The vertical red line indicates a break in the axis in order to illustrate extreme outliers*

Data table for Figure 10

Haemophilia Centre	Patients treated	Patients treated with weight reported	Total Units	Median Units	Median Units/Kg
Aberdeen	11	8	2,892,750	228,000	2,142
Belfast	40	39	9,500,500	217,000	2,707
Birmingham	70	40	12,838,000	165,750	2,276
Bristol	28	7	5,757,000	165,000	1,220
Cambridge	32	31	10,023,000	332,750	3,671
Canterbury	18	14	5,729,000	308,500	2,980
Cardiff	35	23	6,079,353	164,000	1,821
Dundee	12	12	3,142,000	268,000	2,552
Edinburgh	20	18	5,793,250	233,500	3,402
Glasgow	38	38	9,769,000	234,000	3,132
Hammersmith	25	22	8,234,500	350,500	4,931
Leeds	31	12	14,883,000	318,000	5,280
Leicester	18	15	4,089,027	241,000	2,440
Liverpool	29	29	6,153,000	173,000	2,166
Manchester	52	48	11,820,250	235,750	2,937
Newcastle upon Tyne	39	38	12,328,500	281,000	3,680
North Hampshire	53	45	14,129,344	249,000	3,066
Nottingham	26	6	6,251,000	231,750	2,647
Oxford	88	85	20,686,500	209,000	2,477
Royal Free	122	116	38,606,950	317,400	3,880
Royal London	43	27	12,883,410	285,000	4,515
Sheffield	17	16	4,410,000	242,000	3,163
Southampton	12	11	3,236,000	273,750	4,209
St George's	26	26	5,812,000	241,500	2,794
St Thomas'	102	48	38,770,000	348,000	4,259
Centres with <=4 patients meeting criteria	27	17	7,790,000	265,000	2,860
Centres with 5-9 patients meeting criteria	45	26	10,957,750	180,000	2,392

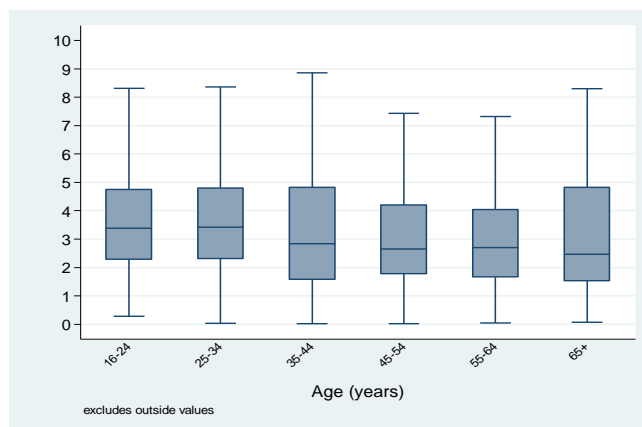
**Figures 11a&b Median factor VIII units used per kilogram body weight per year in Severe Haemophilia A patients without inhibitors by age, 2015/16**

**11a. Paediatrics**



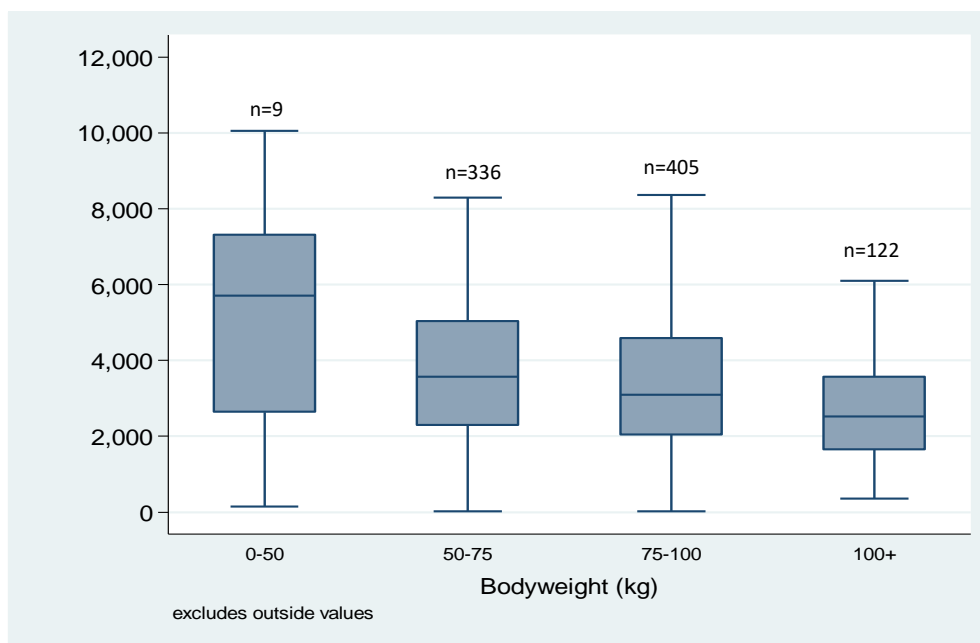
**11b. Adults**

*Weight data are missing for 84/481, including 14/34 aged <12m*



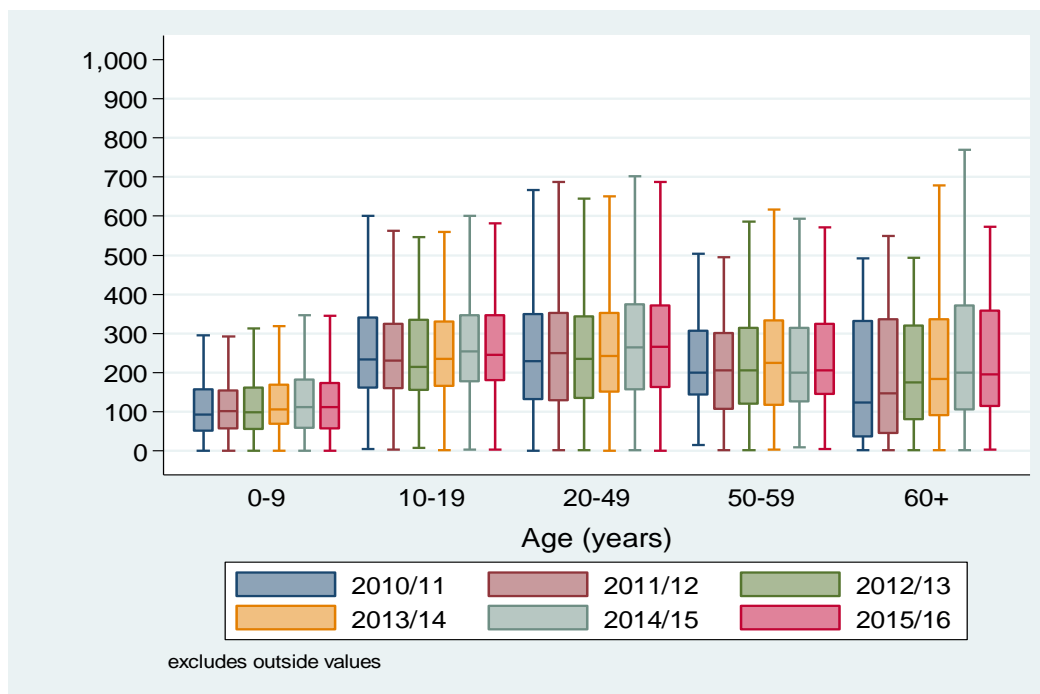
*Weight data are missing for 257/1129*

**Figure 12** Median factor VIII units used per kilogram body weight per year in Severe Haemophilia A patients without inhibitors aged 16 and over, by bodyweight 2015/16

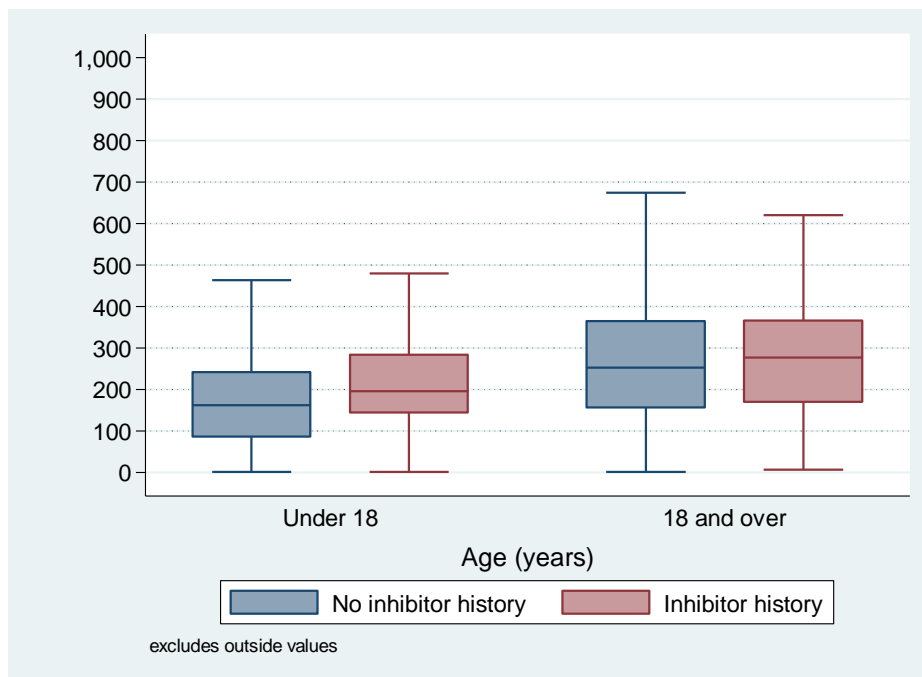


*Weight data are missing for 257/1129 SHA patients known to be aged 16+*

**Figure 13** Median usage of factor VIII: UK patients with severe Haemophilia A without inhibitors broken down by age, 2010/11 – 2015/16



**Figure 14** Severe Haemophilia A patients with no current inhibitor between April 2015 & March 2016: median usage by inhibitor history





**Table 6**      **Products used to treat Haemophilia A (including inhibitors) between April 2015 & March 2016**

Manufacturer	Product	Total Units
Baxalta	Advate	164,292,360
	FEIBA	36,176,573
Bayer	Kogenate	21,489,750
Biotest	Haemoctin	340,000
BPL	FVIII 8Y	256,620
	Optivate	4,715,460
CSL Behring	Haemate P	202,000
	Helixate Nexgen	27,913,250
	Kybernin	6,500
Grifols	Alphanate	142,000
	Fanhdi	25,281,600
Novo Nordisk	NovoSeven (mg)	23,124.00
Octapharma	Nuwiq	5,218,500
	Octanate	3,572,500
Pfizer	BeneFIX	1,000
	ReFacto AF	276,868,501
SOBI/Biogen	Elocta	285,250.00
Various Manufacturers	Investigational Factor VIII	16,147,178

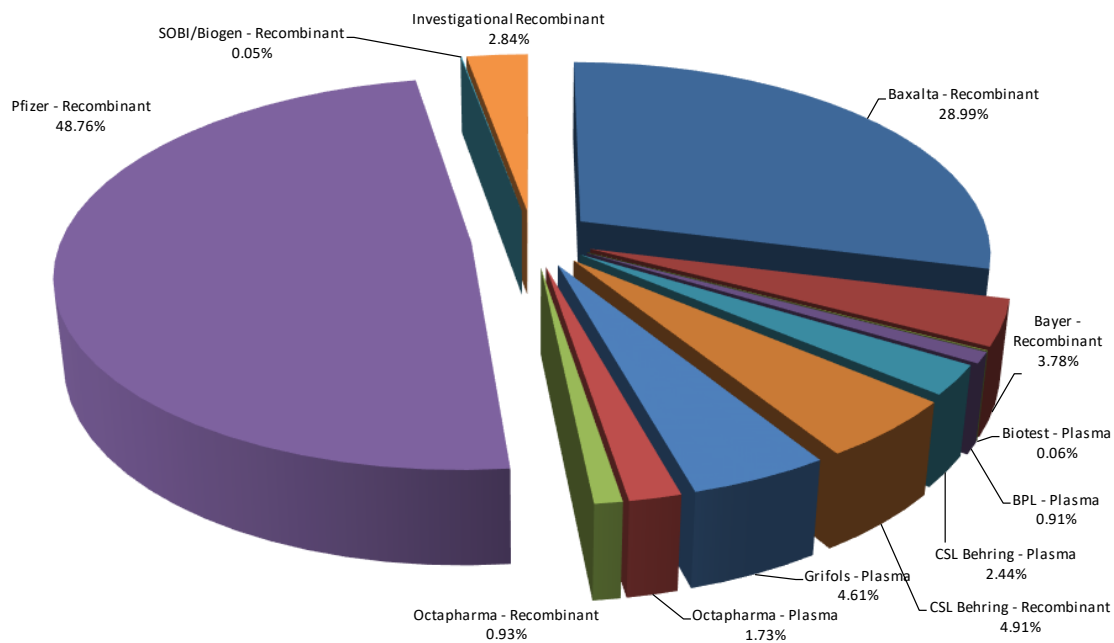
*Units in IU unless otherwise stated*

**Table 7** Factor VIII units used by UK Haemophilia Centres between April 2015 & March 2016, broken down by diagnosis

Coagulation Defect	Patients Treated	Plasma FVIII (IU)	Recombinant FVIII (IU)	Investigational FVIII (IU)	Total FVIII (IU)
Haemophilia A	2,966	34,510,180	494,997,361	16,147,178	545,654,719
Haemophilia A Carrier	41	-	1,070,250	-	1,070,250
Acquired Haemophilia A	7	87,000	48,000	-	135,000
Haemophilia B	1	-	500	-	500
von Willebrand disease	739	19,512,500	43,500	-	19,556,000
Acquired von Willebrand Disease	35	919,800	-	-	919,800
Platelet-type Pseudo von Willebrand D	2	4,500	-	-	4,500
Probable von Willebrand disease	1	2,000	-	-	2,000
Combined V+VIII Deficiency	6	-	35,750	-	35,750
Other combined diagnoses	20	343,500	1,370,000	-	1,713,500
Platelet defects*	1	40,000	-	-	40,000
Miscellaneous*	3	72,500	-	-	72,500
Unclassified*	1	1,000	-	-	1,000
<b>Total</b>	<b>3,823</b>	<b>55,492,980</b>	<b>497,565,361</b>	<b>16,147,178</b>	<b>569,205,519</b>

*N.B: Haemophilia A Carrier includes females with factor VIII deficiency  
Products containing VWF as well as FVIII are reported in FVIII units*

**Figure 15** Market Share of factor VIII concentrates issued by UK Haemophilia Centres during 2015/16



# Haemophilia B

**Table 8 Patients with congenital Haemophilia B registered and treated April 2015 - March 2016**

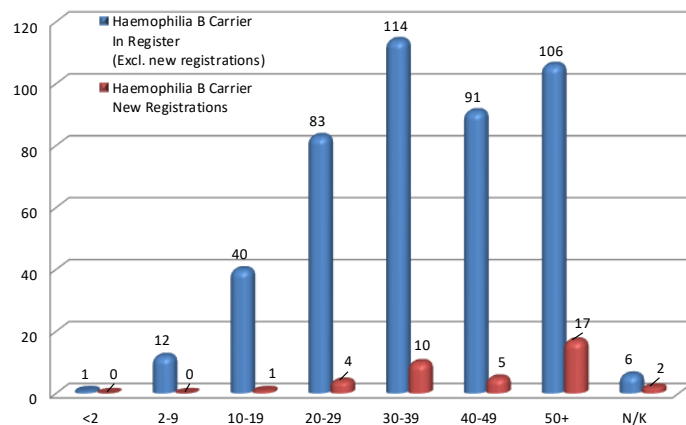
Haemophilia B	Age Range	Factor IX level (iu/dl)					
		< 1	1 - 5	>5 & <40	≥ 40	Unknown	Total
Total In Register (including low-level carriers)	<18 years	111 (0)	86 (0)	141 (44)	29 (24)	2 (2)	369 (70)
	≥18 years	230 (1)	278 (6)	593 (214)	214 (182)	23 (19)	1,338 (422)
	<b>Total</b>	<b>341 (1)</b>	<b>364 (6)</b>	<b>734 (258)</b>	<b>243 (206)</b>	<b>25 (21)</b>	<b>1,707 (492)</b>
New Registrations * (including low-level carriers)	<18 years	7 (0)	4 (0)	10 (6)	3 (3)	1 (1)	25 (10)
	≥18 years	3 (0)	2 (0)	22 (9)	18 (16)	5 (4)	50 (29)
	<b>Total</b>	<b>10 (0)</b>	<b>6 (0)</b>	<b>32 (15)</b>	<b>21 (19)</b>	<b>6 (5)</b>	<b>75 (39)</b>
Treated in year** (including low-level carriers)	<18 years	105 (0)	57 (0)	30 (1)	1 (1)	- (0)	193 (2)
	≥18 years	206 (0)	142 (1)	135 (37)	5 (3)	1 (0)	489 (41)
	<b>Total</b>	<b>311 (0)</b>	<b>199 (1)</b>	<b>165 (38)</b>	<b>6 (4)</b>	<b>1 (0)</b>	<b>682 (43)</b>

\* New registrations are a subset of the 'In Register' numbers

\*\* Treated includes patients 'In Register' and 'New Registrations'

The numbers in brackets are females

**Figure 16** Carriers of Haemophilia B currently registered and newly registered, by baseline factor IX level



Diagnosis	Factor VIII level (iu/dl)								Grand Total
	<2	2-9	10-19	20-29	30-39	40-49	50+	N/K	
Haemophilia B Carrier In Register (Excl. new registrations)	1	12	40	83	114	91	106	6	453
Haemophilia B Carrier New Registrations	0	0	1	4	10	5	17	2	39
	1	12	41	87	124	96	123	8	492

*N.B:*

*Haemophilia B Carrier includes:*

*Females registered by their Haemophilia Centre as Females with FIX deficiency*

*Females registered by their Haemophilia Centre as Haemophilia B*

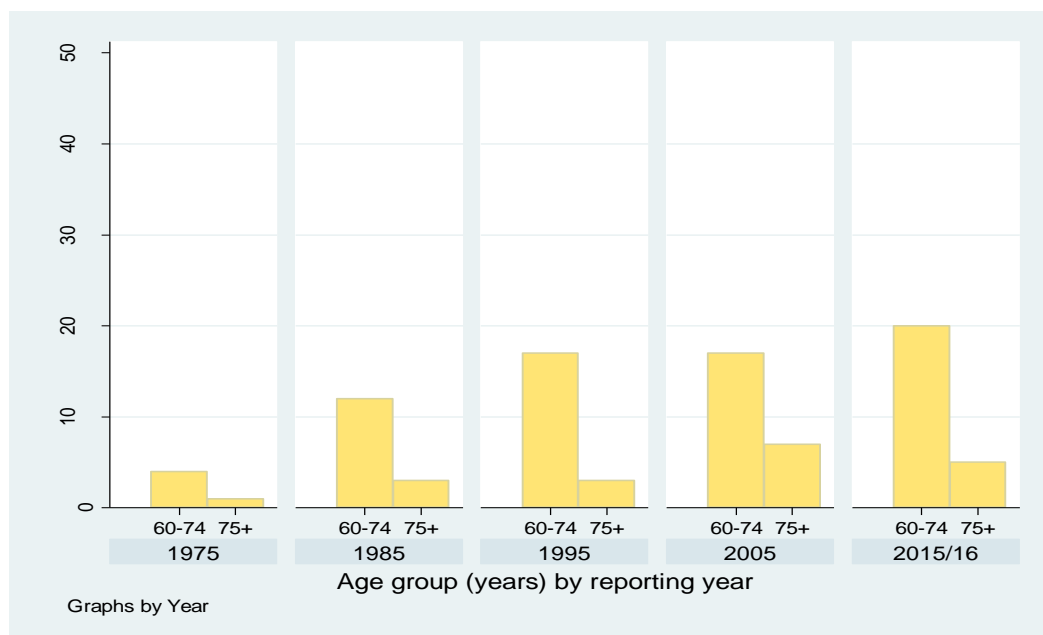
*FIX Leyden carriers*

**Table 9** New registrations of Haemophilia B between April 2015 & March 2016, by age at mid-year and severity

Coagulation Defect	Age (years)	Number of Patients (Factor IX level (iu/dl))					Total
		< 1	1 - 5	>5 & <40	≥ 40	Unknown	
Haemophilia B	0 : 4	6	3	1	0	0	10
	5 : 9	1	1	1	0	0	3
	10 : 19	0	0	4	0	0	4
	20 : 29	3	0	4	1	1	9
	30 : 39	0	0	4	0	0	4
	40 : 49	0	1	2	0	0	3
	50 : 59	0	1	0	1	0	2
	60 : 69	0	0	0	0	0	0
	70 +	0	0	1	0	0	1
Total		10	6	17	2	1	36

*N.B: Haemophilia B includes patients with FIX Leyden*

**Figure 17** Trend in numbers of Severe Haemophilia B patients aged 60 years and above, 1975 - 2015/16 (including HIV +ve pts)





**Figure 18** Trend in numbers of non-Severe Haemophilia B patients aged 60 years and above, 1975 - 2015/16 (including HIV +ve pts)

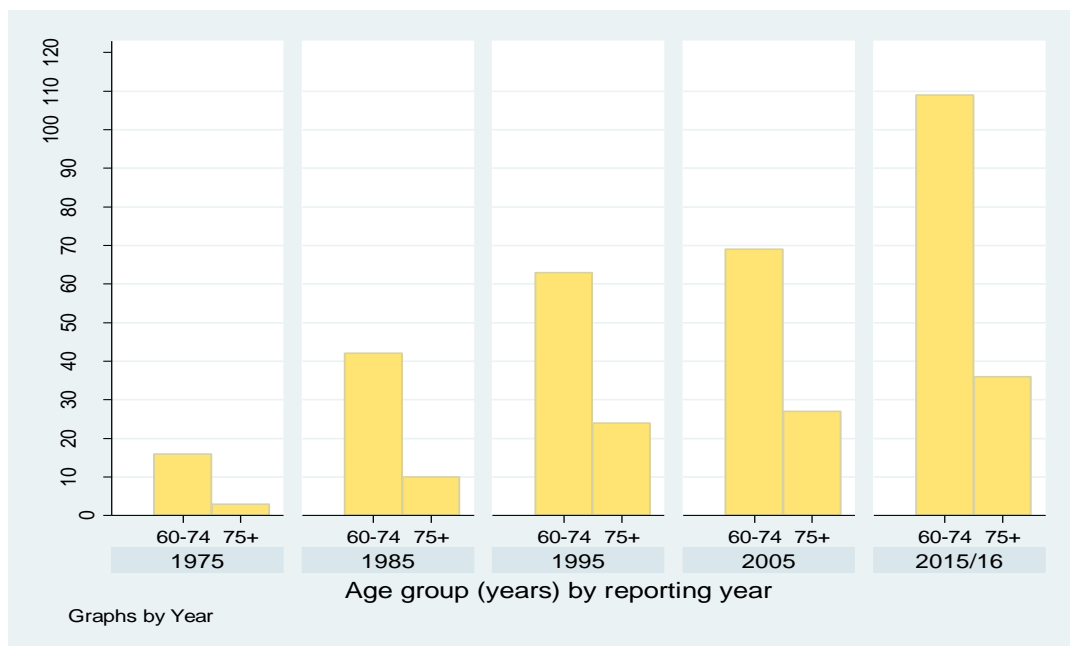
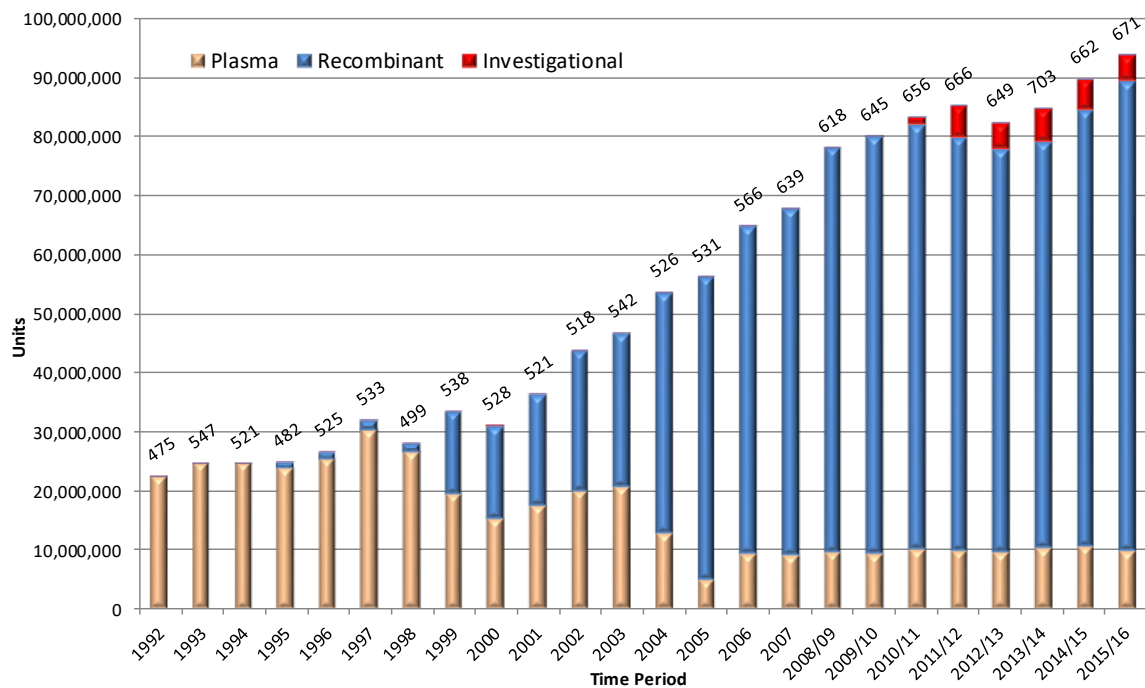


Figure 19

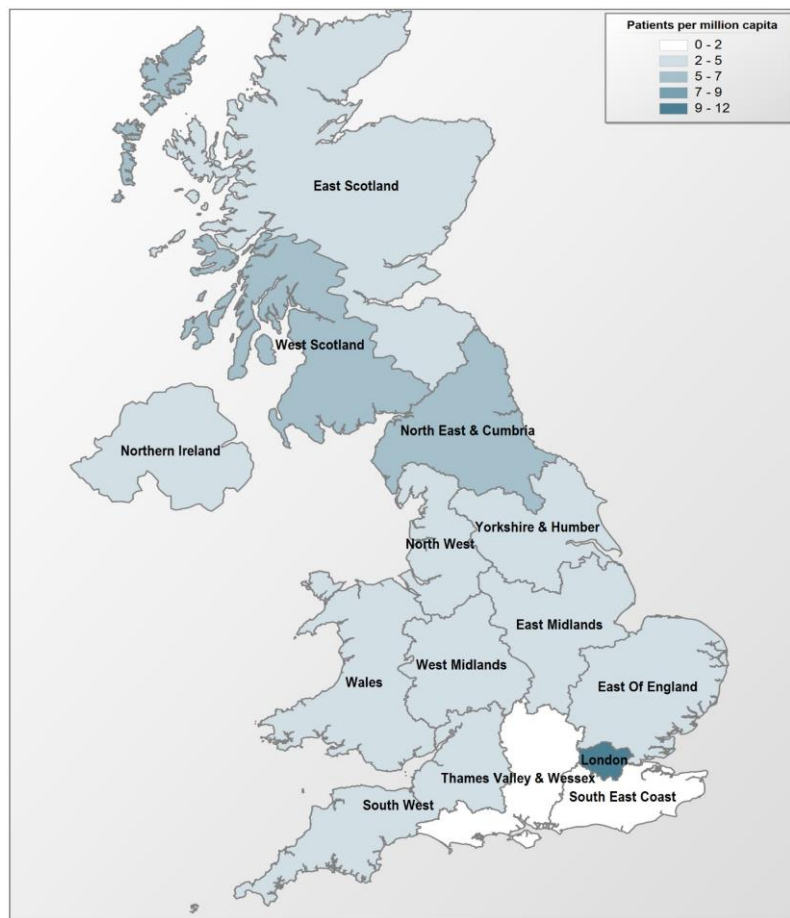
Factor IX units used by UK Haemophilia Centres 1992- 2015/16 all diagnoses



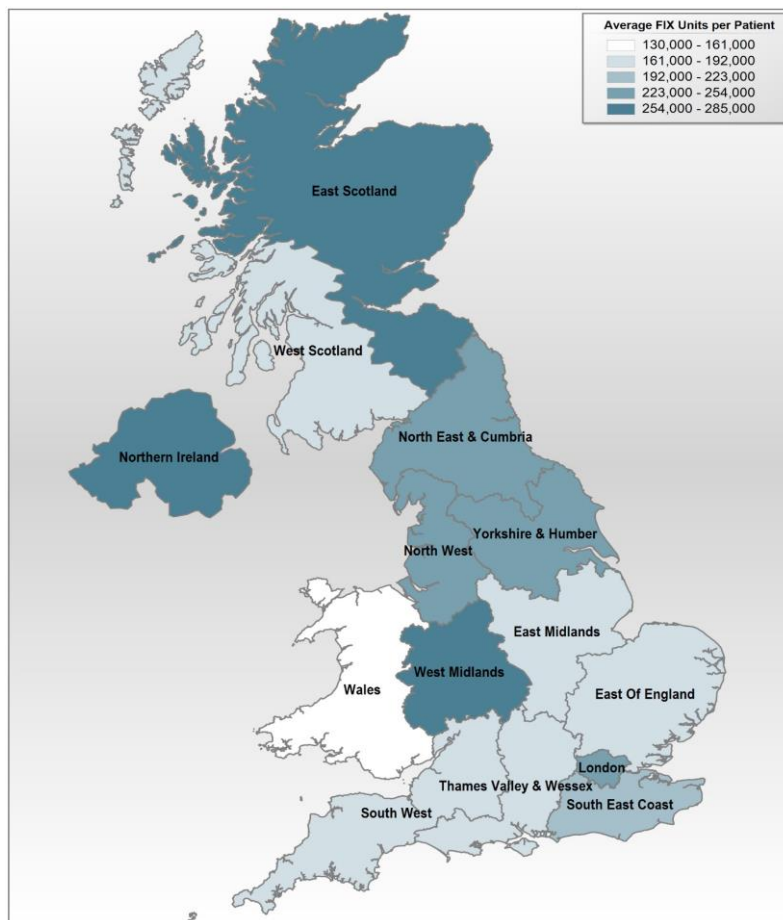
N.B: Data for St Thomas' were not submitted 1996-2006

Figure 20

Number of Severe Haemophilia B patients treated per million capita



**Figure 21** Factor IX Usage (including inhibitor) divided by number of Severe Haemophilia B patients by commissioning region



**Table 10** Factor IX usage by region for patients with Severe Haemophilia B (incl. treatment for inhibitors)

Region	General Population	Patients treated (n) Severe Haemophilia B	Total FIX Units (IU) Severe Haemophilia B	Mean Usage Severe Haemophilia B	FIX Units Per Capita Severe Haemophilia B
Birmingham & Black Country		20	5,448,000	272,400	
Cumbria, Northumberland & Tyne and Wear		16	3,960,000	247,500	
Cheshire, Warrington & Wirral		25	6,089,920	243,597	
London		92	21,424,870	232,879	
South Yorkshire & Bassetlaw		21	4,780,722	227,653	
Surrey & Sussex		7	1,483,500	211,929	
Bristol, North Somerset & South Gloucestershire		14	2,659,500	189,964	
Wessex		37	6,728,870	181,861	
Leicestershire & Lincolnshire		15	2,562,000	170,800	
East Anglia		23	3,919,250	170,402	
<b>England</b>	<b>54,786,300</b>	<b>259</b>	<b>59,056,632</b>	<b>228,018</b>	<b>1.08</b>
Northern Ireland	1,851,600	8	2,231,100	278,888	1.20
Scotland East	3,021,290	9	2,502,000	278,000	0.83
Scotland West	2,351,710	14	2,261,250	161,518	0.96
Wales	3,099,100	10	1,392,553	139,255	0.45
<b>United Kingdom</b>	<b>65,110,000</b>	<b>300</b>	<b>67,443,535</b>	<b>224,812</b>	<b>1.04</b>

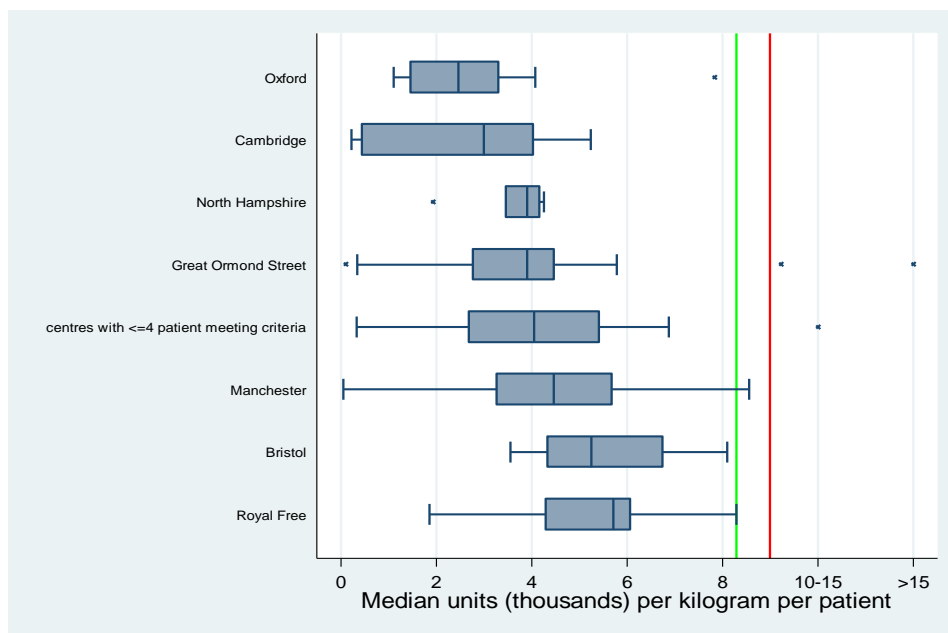
English regions ranked by mean usage

**Source accreditation:**

England, Northern Ireland & Wales: Mid-2015 Population estimate: Office for National Statistics licensed under the Open Government Licence v.3.0.

Scotland: Mid-2015 Population Estimates supplied by National Records of Scotland under the Open Government Licence v3.0 © Crown Copyright 2016

**Figure 22** Annual FIX usage 2015/16 in Severe Haemophilia B patients aged under 18 years with no current inhibitor, by centre, ranked by median



Indicated outside values show where a patient's usage is more than 1.5 times the IQR from the nearest quartile at centre level

The vertical green line indicates the 95<sup>th</sup> percentile of units, regardless of centre (8,291 IU/kg)

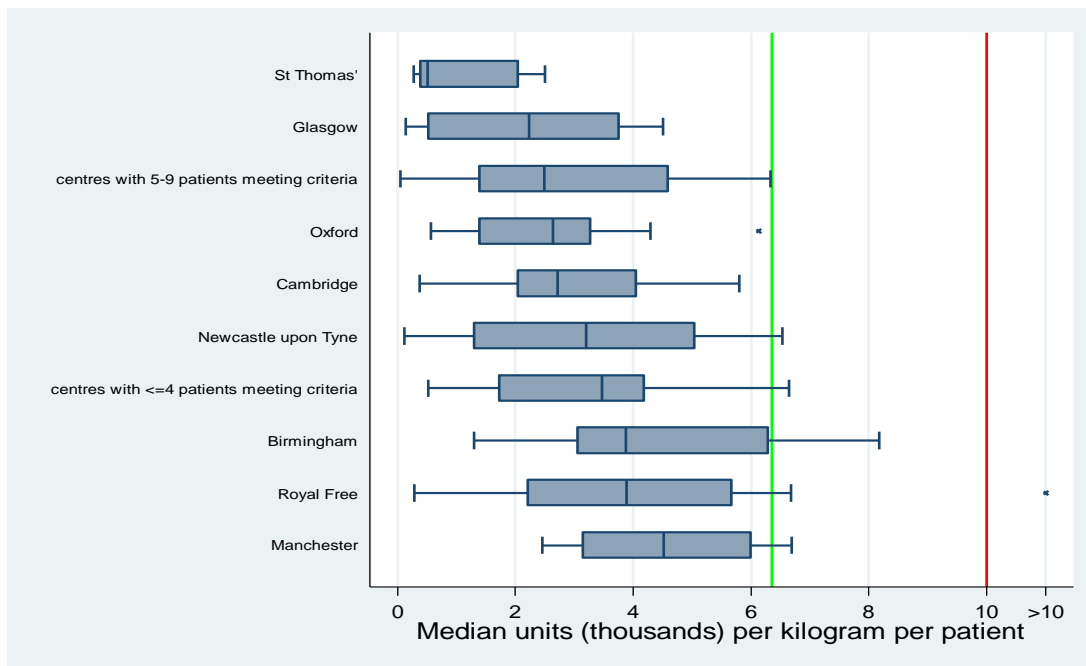
The vertical red line indicates a break in the axis in order to illustrate extreme outliers

Patients treated at more than 1 centre during the year have all their usage reported against the centre where they received the most units

Data table for Figure 22

Haemophilia Centre	Patients treated	Patients treated with weight reported	Total Units	Median Units	Median Units/Kg
Bristol	5	4	632,500	117,500	5,253
Cambridge	6	6	563,000	62,375	2,994
Great Ormond Street	23	23	2,922,314	123,250	3,909
Manchester	9	9	1,375,500	140,000	4,464
North Hampshire	5	5	547,250	109,000	3,902
Oxford	9	9	1,132,500	64,500	2,462
Royal Free	5	5	1,673,000	344,000	5,707
Centres with <=4 patients meeting criteria	39	35	5,743,152	96,000	4,058

**Figure 23** Annual FIX usage 2015/16 in Severe Haemophilia B patients aged 18 years or more with no current inhibitor, by centre, ranked by median usage



Indicated outside values show where a patient's usage is more than 1.5 times the IQR from the nearest quartile at centre level

The vertical green line indicates the 95<sup>th</sup> percentile of units, regardless of centre 6,356 IU/kg)

The vertical red line indicates a break in the axis in order to illustrate extreme outliers

Patients treated at more than 1 centre during the year have all their usage reported against the centre where they received the most units



Data table for Figure 23

Treatment Year	Haemophilia A <1		Haemophilia A 1 - 5		Haemophilia A >5 & <40		Haemophilia A ≥ 40	
	n	% difference since 2007	n	% difference since 2007	n	% difference since 2007	n	% difference since 2007
2007	1405	100.0	587	100.0	583	100.0	26	100.0
2008/09	1454	103.5	587	100.0	561	96.2	22	84.6
2009/10	1475	105.0	569	96.9	617	105.8	26	100.0
2010/11	1511	107.5	578	98.5	641	109.9	26	100.0
2011/12	1550	110.3	594	101.2	650	111.5	20	76.9
2012/13	1562	111.2	572	97.4	647	111.0	19	73.1
2013/14	1623	115.5	603	102.7	652	111.8	19	73.1
2014/15	1663	118.4	603	102.7	648	111.1	21	80.8
2015/16	1710	121.7	605	103.1	668	114.6	21	80.8

**Table 11**      **Products used to treat Haemophilia B (including inhibitors) between April 2015 & March 2016**

Manufacturer	Product	Total Units
Baxalta	FEIBA	2,204,000
BPL	Replenine	2,686,610
CSL Behring	Mononine	540,000
Grifols	Alphanine	6,338,600
Novo Nordisk	NovoSeven (mg)	8,295
Pfizer	BeneFIX	79,040,670
	ReFacto AF	500
SOBI/Biogen	ALPROLIX	54,000
Various Manufacturers	Investigational Factor IX	4,361,406
<b>Total</b>		<b>95,234,081</b>

*Units in IU unless otherwise stated*

**Table 12** Factor IX units issued by UK Haemophilia Centres between April 2015 & March 2016, broken down by diagnosis table for Figures 6a, 6b & 6c

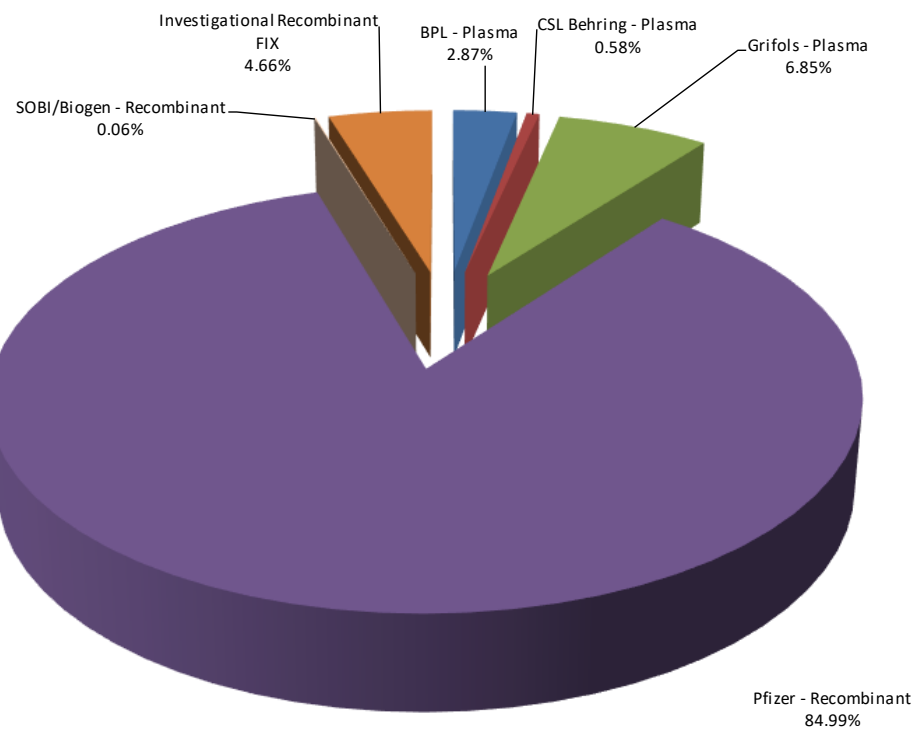
Coagulation Defect	Patients Treated	Plasma FIX (IU)	Recombinant FIX (IU)	Investigational FIX (IU)	Total FIX (IU)
Haemophilia A	1	-	1,000	-	1,000
Haemophilia B	628	9,565,210	79,308,170	4,361,406	93,234,786
Haemophilia B Carrier	41	78,000	338,500	-	416,500
Combined Diagnoses	1	-	5,000	-	5,000
<b>Total</b>	<b>671</b>	<b>9,643,210</b>	<b>79,652,670</b>	<b>4,361,406</b>	<b>93,657,286</b>

*N.B:*

*Haemophilia B includes patients with FIX Leyden*

*Haemophilia B Carrier includes Females with FIX deficiency and FIX Leyden carriers*

**Figure 24**    **Market Share of factor IX concentrates issued by UK Haemophilia Centres during 2015/16**



# Von Willebrand Disease

**Table 13 Patients with von Willebrand Disease registered and treated April 2015 - March 2016**

von Willebrand disease	<18 years (VWD Activity iu/dl)					≥18 years (VWD Activity iu/dl)					Total	Treated
	<10	10 - 29	≥30	N/K	Sub Total	<10	10 - 29	≥30	N/K	Sub Total		
Males												
Type 1	26	155	285	15	481	81	344	641	105	1,171	1,652	76
Type 2A	33	34	8	1	76	90	70	38	3	201	277	65
Type 2B	6	10	3	0	19	12	30	15	2	59	78	17
Type 2M	19	5	5	0	29	26	27	10	3	66	95	12
Type 2N	0	0	2	0	2	3	2	22	6	33	35	1
Type 2 Unspecified	4	8	6	0	18	11	12	8	1	32	50	5
Type 3	32				32	54				54	86	61
Type Unreported	53	114	244	10	421	158	335	611	58	1,162	1,583	96
Sub Total Males											3,856	333
Females												
Type 1	25	105	238	6	374	126	532	1,737	215	2,610	2,984	101
Type 2A	29	22	10	0	61	111	99	66	7	283	344	68
Type 2B	3	5	10	0	18	14	43	38	1	96	114	22
Type 2M	7	20	8	1	36	48	51	20	7	126	162	28
Type 2N	0	1	1	0	2	6	7	48	5	66	68	6
Type 2 Unspecified	4	1	0	0	5	18	21	20	2	61	66	3
Type 3	18				18	50				50	68	42
Type Unreported	50	137	219	10	416	208	583	1,557	172	2,520	2,936	141
Sub Total Females											6,742	411
Grand Total - Males and Females											10,598	744

**Table 14** New Registrations of von Willebrand Disease between April 2015 - March 2016, by age at mid-year, severity, and gender

von Willebrand disease	<18 years (VWD Activity iu/dl)					≥18 years (VWD Activity iu/dl)					Total
	<10	10 - 29	≥30	N/K	Sub Total	<10	10 - 29	≥30	N/K	Sub Total	
Males											
Type 1	3	9	12	0	24	0	7	9	0	16	40
Type 2A	1	4	0	0	5	0	3	2	0	5	10
Type 2B	0	0	0	0	0	1	0	1	0	2	2
Type 2M	1	0	0	0	1	0	1	0	0	1	2
Type 2N	0	0	0	0	0	0	0	2	1	3	3
Type 2 Unspecified	0	0	0	0	0	0	0	0	0	0	0
Type 3	0				0	1				1	1
Type Unreported	8	19	30	0	57	3	13	8	3	27	84
Sub Total Males											142
Females											
Type 1	1	10	10	0	21	0	14	54	1	69	90
Type 2A	2	5	0	0	7	2	1	0	0	3	10
Type 2B	0	0	0	0	0	0	1	1	0	2	2
Type 2M	0	2	1	0	3	2	1	0	0	3	6
Type 2N	0	0	0	0	0	0	0	2	0	2	2
Type 2 Unspecified	0	0	0	0	0	0	0	0	0	0	0
Type 3	1				1	1				1	2
Type Unreported	5	24	25	1	55	4	16	67	3	90	145
Sub Total Females											257
Grand Total - Males and Females											399

**Table 15**      **Concentrates used to treat von Willebrand Disease (including inhibitors) between April 2015 & March 2016**

Manufacturer	Product	Total Units
Baxalta	Advate	14,500
BPL	FVIII 8Y	40,000
CSL Behring	Haemate P	7,625,150
	Voncento	5,353,350
Grifols	Alphanate	780,000
	Fanhdi	4,000
LFB Biomedicaments	Willfact /Wilfactin	555,000
Novo Nordisk	NovoSeven (mg)	1,956
Octapharma	Octaplex	500
	Wilate	5,710,000
Pfizer	ReFacto AF	29,000
	Investigational rVWF	*

*\*Anonymised for confidentiality purposes*

*Units in IU unless otherwise stated*

*Products containing VWF and FVIII are reported in FVIII units*



# Inhibitors: Congenital and Acquired Haemophilia A, Haemophilia B and von Willebrand Disease

**Table 16 Inhibitors by Disease Severity - Haemophilia A, Haemophilia B & von Willebrand Disease**

Coagulation Defect	Severity / Subtype	In Register *	Inhibitors			
			Newly Reported n (%)	Ongoing n (%)	Current n (%)	Historical n (%)
Haemophilia A	< 1 iu/dl	1,845	19 (1.0)	145 (7.9)	164 (8.9)	412 (22.3)
	1 - 5 iu/dl	882	4 (0.5)	38 (4.3)	42 (4.8)	96 (10.9)
	>5 - <40 iu/dl	3,203	6 (0.2)	18 (0.6)	24 (0.7)	63 (2.0)
	<b>Total</b>	<b>5,930</b>	<b>29 (0.5)</b>	<b>201 (3.4)</b>	<b>230 (3.9)</b>	<b>571 (9.6)</b>
Haemophilia B	< 1 iu/dl	340	2 (0.6)	10 (2.9)	12 (3.5)	17 (5.0)
	1 - 5 iu/dl	358	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
	>5 - <40 iu/dl	516	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
	Unknown	1	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
	<b>Total</b>	<b>1,215</b>	<b>2 (0.2)</b>	<b>10 (0.8)</b>	<b>12 (1.0)</b>	<b>17 (1.4)</b>
von Willebrand disease	Type 1	4,626	0 (0.0)	0 (0.0)	0 (0.0)	1 (0.0)
	Type 3	154	0 (0.0)	6 (3.9) **	6 (3.9)	6 (3.9)
	Others	5,819	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
	<b>Total</b>	<b>10,599</b>	<b>0 (0.0)</b>	<b>6 (0.1)</b>	<b>6 (0.1)</b>	<b>7 (0.1)</b>

\* Including patients not regularly treated

\*\* two of these patients had no treatment in the reporting year

N.B: Haemophilia B includes patients with FIX Leyden

New=new in year

Ongoing=inhibitor in year but not new in year

Current=Sum of New + Ongoing

Historical=history of inhibitor, but not current

**Table 17 Annual concentrate usage by patients with *congenital* bleeding disorders who were inhibitor positive at any time within 2015/16**

Manufacturer	Product	Haemophilia A	Haemophilia B	von Willebrand Disease	F.VII deficiency	F.XI deficiency	Combined Diagnoses
Baxalta	Advate	17,572,750	-	-	-	-	-
	FEIBA	36,174,573	2,204,000	-	-	-	932,500
Bayer	Kogenate	5,596,000	-	-	-	-	-
BPL	FVIII 8Y	256,620	-	-	-	-	-
	FXI	-	-	-	-	2,785	-
	Optivate	3,934,460	-	-	-	-	-
CSL Behring	Haemate P	201,000	-	197,000	-	-	-
	Helixate Nexgen	1,940,250	-	-	-	-	-
	Voncento	-	-	34,500	-	-	-
Grifols	Fanhdri	12,885,600	-	-	-	-	-
Novo Nordisk	NovoSeven (mg)	23,020	8,295	1,481	573	8	892
Octapharma	Nuwiq	121,250	-	-	-	-	-
	Octanate	2,361,000	-	-	-	-	-
Pfizer	Benefix	-	2,000	-	-	-	-
	ReFacto AF	12,014,500	-	10,000	-	-	-
Various Manufacturers	Investigational Factor VIII	143,681	-	-	-	-	-

*Units in IU unless otherwise stated*

**Table 18**      **Concentrates used to treat Acquired Inhibitors**

Manufacturer	Product	Acquired Haemophilia A	Acquired von Willebrands	Acquired F.VII Deficiency	Acquired F.XIII Deficiency
Baxalta	FEIBA	5,263,050	-	18,000	-
Bayer	Optivate	32,000	-	-	-
CSL Behring	Fibrogammin P	-	-	-	9,000
	Haemate P	-	263,500	-	-
	Voncento	-	108,300	-	-
Grifols	Fanhdi	55,000	-	-	-
IDEC Pharmaceuticals	Rituximab	9,315	-	-	-
Novo Nordisk	NovoSeven (mg)	2,246	-	-	-
Octapharma	Wilate	-	548,000	-	-
Pfizer	ReFacto AF	48,000	-	-	-

*Units in IU unless otherwise stated*

**Table 19** FEIBA® usage: breakdown by diagnosis

Coagulation Defect	Number of Patients	Total (u)
Haemophilia A	98	36,176,573
Haemophilia B	6	2,204,000
Acquired Haemophilia A	88	5,263,050
F.XI Deficiency	1	2,000
Acquired Deficiency (other)	2	18,000
Combined Diagnoses	2	932,500
	<b>197</b>	<b>44,596,123</b>

**Table 20** NovoSeven® usage: breakdown by diagnosis

Coagulation Defect	Number of Patients	Total (mg)
Haemophilia A	77	23,124
Haemophilia B	10	8,295
von Willebrand disease	3	1,956
Acquired Haemophilia A	29	2,246
F.V deficiency	2	10
F.VII deficiency	52	2,174
F.XI Deficiency	2	16
Combined Diagnoses	3	894
Bernard Soulier	8	378
Glanzmann's Thrombasthenia	48	1,404
Platelet defects	3	37
Unclassified	2	30
	<b>239</b>	<b>40,565</b>

# Rarer Bleeding Disorders

**Table 21 Patients with rarer types of bleeding disorders registered and treated April 2015 & March 2016**

Coagulation Defect	Number of Patients in Register			Treated	% Treated
	Total	Males	Females		
Probable von Willebrand disease	185	48	137	11	5.95%
Platelet-type Pseudo von Willebrand Disease	19	9	10	2	10.53%
F.V deficiency	200	79	121	12	6.00%
F.VII deficiency	1,148	558	590	54	4.70%
F.X deficiency	241	100	141	35	14.52%
F.XI Deficiency	2,853	1,202	1,651	85	2.98%
F.XIII Deficiency	66	37	29	53	80.30%
Combined II+VII+IX+X Deficiency	4	1	3	0	0.00%
Combined V+VIII Deficiency	27	13	14	6	22.22%
Other combined diagnoses	321	144	177	39	12.15%
Prothrombin Deficiency	14	5	9	3	21.43%
Afibrinogenemia	12	8	4	10	83.33%
Dysfibrinogenemia	426	166	260	25	5.87%
Hypofibrinogenemia	115	47	68	6	5.22%
Hypodysfibrinogenemia	1	0	1	1	100.00%
Fibrinogen Deficiency	43	21	22	9	20.93%
Acquired Haemophilia A	475	236	239	102	21.47%
Acquired Haemophilia B	2	2	0	0	0.00%
Acquired von Willebrands	117	62	55	36	30.77%
Acquired Prothrombin Deficiency	5	3	2	0	0.00%
Acquired F.XIII Deficiency	3	2	1	1	33.33%
Acquired F.V Deficiency	4	0	4	0	0.00%
Acquired Deficiency (other)	6	5	1	2	33.33%
Glanzmann's Thrombasthenia	125	53	72	52	41.60%
Bernard Soulier	85	42	43	9	10.59%
Other platelet defects	2,222	695	1,527	115	5.18%
Miscellaneous	248	66	182	11	4.44%
Unclassified bleeding disorder	359	56	303	14	3.90%
Haemophilia A with Liver Transplant	10	10	0	1	10.00%
Haemophilia B with Liver Transplant	3	3	0	0	0.00%
von Willebrand with Liver Transplant	2	2	0	0	0.00%
<b>Total</b>	<b>9,341</b>	<b>3,675</b>	<b>5,666</b>	<b>694</b>	<b>7.43%</b>



**Table 22** Patients with selected rarer bleeding disorders registered and treated April 2015 - March 2016, 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	46	8	154	4	0	0	200	12
F.VII deficiency	123	23	1025	31	0	0	1148	54
F.X deficiency	37	25	204	10	0	0	241	35
F.XI Deficiency	225	24	2629	61	0	0	2854	85
<b>Total</b>	<b>431</b>	<b>80</b>	<b>4,012</b>	<b>106</b>	<b>-</b>	<b>-</b>	<b>4,443</b>	<b>186</b>

Coagulation Defect	<2		≥2		N/K		Total	
	In Reg	Treated	In Reg	Treated	In Reg	Treated	In Reg	Treated
F.XIII Deficiency	36	33	30	20	0	0	66	53
<b>Total</b>	<b>36</b>	<b>33</b>	<b>30</b>	<b>20</b>	<b>-</b>	<b>-</b>	<b>66</b>	<b>53</b>

**Table 23** New registrations of rarer bleeding disorders between April 2015 & March 2016  
showing their coagulation defect and gender

Coagulation Defect	Male	Female	Total
Haemophilia A with Liver Transplant	3	0	3
Haemophilia B with Liver Transplant	1	0	1
Platelet-type Pseudo von Willebrand Disease	3	4	7
Probable von Willebrand disease	7	20	27
F.V deficiency	1	7	8
F.VII deficiency	61	73	134
F.X deficiency	4	10	14
F.XI Deficiency	102	134	236
F.XIII Deficiency	0	1	1
Afibrinogenemia	2	2	4
Dysfibrinogenemia	28	59	87
Hypofibrinogenemia	26	33	59
Fibrinogen Deficiency	0	1	1
Prothrombin Deficiency	0	2	2
Acquired Haemophilia A	63	43	106
Acquired F.V deficiency	0	1	1
Acquired F.XIII Deficiency	1	0	1
Acquired Prothrombin Deficiency	1	1	2
Acquired von Willebrands	15	9	24
Glanzmann's Thrombasthenia	3	4	7
Bernard Soulier	4	3	7
Other platelet defects	85	199	284
Other combined diagnoses	17	19	36
Miscellaneous	6	31	37
Unclassified bleeding disorder	16	76	92
<b>Total</b>	<b>449</b>	<b>732</b>	<b>1,181</b>

**Table 24 Concentrates used to treat Rarer Bleeding Disorders between April 2015 & March 2016**

Manufacturer	Product	F.V deficiency		F.VII deficiency		F.IX Leyden		F.IX Leyden Carrier		F.X deficiency		F.XI Deficiency		F.XIII Deficiency	
		Pts treated	Units	Pts treated	Units	Pts treated	Units	Pts treated	Units	Pts treated	Units	Pts treated	Units	Pts treated	Units
Baxalta	FEIBA	-	-	-	-	-	-	-	-	-	-	1	2,000	-	-
	FVII	-	-	2	105,600	-	-	-	-	-	-	-	-	-	-
BPL	FX	-	-	-	-	-	-	-	-	15	820,615	-	-	-	-
	FXI	-	-	-	-	-	-	-	-	-	-	62	160,700	-	-
CSL Behring	Beriplex	-	-	-	-	-	-	-	-	15	692,000	-	-	-	-
	Fibrogammin P	-	-	-	-	-	-	-	-	-	-	-	-	52	780,914
	FXIII	-	-	-	-	-	-	-	-	-	-	-	-	1	1,250
	Riastap (mg)	-	-	-	-	-	-	-	-	-	-	1	2	-	-
LFB Biomedicaments	Hemoleven	-	-	-	-	-	-	-	-	-	-	4	8,000	-	-
Novo Nordisk	NovoSeven (mg)	2	10	53	2,174	-	-	-	-	-	-	2	16	-	-
	NovoThirteen	-	-	-	-	-	-	-	-	-	-	-	-	1	30,000
Octapharma	Octaplas (units)	9	451	-	-	-	-	-	-	-	-	16	1,908	-	-
	Octaplex	-	-	-	-	-	-	-	-	12	486,000	-	-	-	-
Pfizer	BeneFIX	-	-	-	-	7	213,500	1	17,500	-	-	-	-	-	-

*Units in IU unless otherwise stated*

# Adverse Events

**Table 25**      **Adverse Events reported between April 2015 & March 2016**

Adverse Event	Number of Patients	Number of Events
Allergy Event	5	5
Infection Event	0	0
Inhibitor Event	22	22
Intracranial Haemorrhage	5	5
Malignancy Event	13	13
Other Event	0	0
Poor Efficacy Event	1	1
Thrombotic Event	2	2
<b>Total</b>	<b>48</b>	<b>48</b>

*See table 16 for breakdown of inhibitors by disease severity in Haemophilia A, B and von Willebrand disease*

**Table 26** Summary of patients 'at risk' of vCJD for public health purposes who received UK sourced plasma products as reported by Centres

Summary table of 'at risk' bleeding disorder patients who received UK sourced plasma products				
		Implicated batches	Non-implicated batches	Combined
Current status of 'at risk' patients	Alive	691	2593	3284
	Dead	118	623	741
	Total	809	3216	4025
Genotype	MM	-	-	5
	MV	-	-	4
	VV	-	-	1
	Not known	809	3216	
Sex	M	769	2639	3408
	F	40	577	617
Current age band of living 'at risk' patients	0-19	0	28	28
	20-39	326	825	1151
	40-59	267	1058	1325
	60-79	90	570	660
	80+	8	110	118
	Not known	0	2	2

*These data were last updated on 30/06/2016.*

# Morbidity and Mortality

**Table 27 Causes of death in patients with Haemophilia A, Haemophilia B and all Carriers of Haemophilia A & B between April 2015 & March 2016**

Cause of Death	Severity (factor level iu/dl)					Total
	< 1	1 - 5	>5 & <40	≥ 40	Unknown	
Amyloidosis	0	0	1	0	0	1
ARDS	1	0	0	0	0	1
Carcinoma	0	1	4	0	1	6
Cerebral haemorrhage	0	0	1	0	0	1
Haemorrhage (Misc)	3	0	1	0	0	4
Hepatocellular Carcinoma	0	0	1	0	0	1
Infection (Bacterial)	0	0	3	1	0	4
Ischaemic Heart Disease	1	1	2	0	0	4
Leukaemia	0	0	1	0	0	1
Liver Failure	5	1	0	0	0	6
Parkinson's Disease	0	0	2	0	0	2
Post-operative complications	0	1	0	0	0	1
Stroke (Unknown)	0	0	1	0	0	1
Unknown	2	5	20	4	0	31
<b>Total</b>	<b>12</b>	<b>9</b>	<b>37</b>	<b>5</b>	<b>1</b>	<b>64</b>

*N.B: Includes FIX Leyden and FIX Leyden carriers*



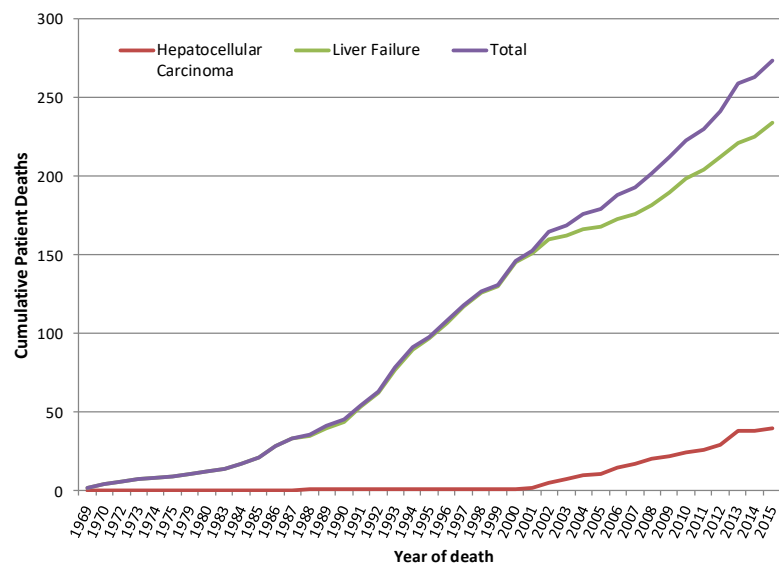
**Table 28 Causes of death in other coagulation defects**

Coagulation Defect	Cause of Death	Total
Haemophilia A with Liver Transplant	Haemorrhage (Misc)	1
	Unknown	1
F.V deficiency	ARDS	1
	Unknown	1
F.VII deficiency	Carcinoma	2
	Ischaemic Heart Disease	1
	Liver Failure	1
	Lymphoproliferative Malignancy	1
	Unknown	5
F.X deficiency	Motor Neurone Disease	1
	Unknown	1
F.XI Deficiency	Carcinoma	4
	COAD	1
	Infection (Bacterial)	3
	Ischaemic Heart Disease	1
	Unknown	16
Combined diagnoses	Ischaemic Heart Disease	1
	Unknown	1
Combined V+VIII Deficiency	Renal Failure	1
Acquired Haemophilia A	AIDS	1
	ARDS	1
	Carcinoma	4
	Cerebral haemorrhage	2
	COAD	2
	Infection (Bacterial)	4
	Intestinal Obstruction	1
	Ischaemic Heart Disease	3
	Liver Failure	1
	Senility/Alzheimer's disease	1
	Unknown	51
Acquired F.V deficiency	Unknown	1
Acquired F.XIII Deficiency	Unknown	1
Acquired von Willebrand Disease	Infection (Bacterial)	1
	Lymphoproliferative Malignancy	1
	Unknown	3
Fibrinogen Deficiency	Unknown	3
Dysfibrinogenemia	Carcinoma	1
	Intestinal obstruction	1
	Unknown	3

**Table 28 Causes of death in other coagulation defects (continued)**

Coagulation Defect	Cause of Death	Total
Hypofibrinogenemia	Unknown	2
Platelet defects	Carcinoma	3
	Ischaemic Heart Disease	1
	Liver Failure	1
	Stroke (Unknown)	2
	Unknown	7
Miscellaneous	Unknown	1
Unclassified	Carcinoma	1
	Cerebral haemorrhage	1
	Unknown	1
von Willebrand disease	AIDS	1
	ARDS	1
	Carcinoma	10
	Cerebral haemorrhage	2
	Dissecting Aortic Aneurysm	1
	Haemorrhage (Misc)	1
	Infection (Bacterial)	6
	Ischaemic Heart Disease	2
	Unknown	38
<b>Total</b>		<b>212</b>

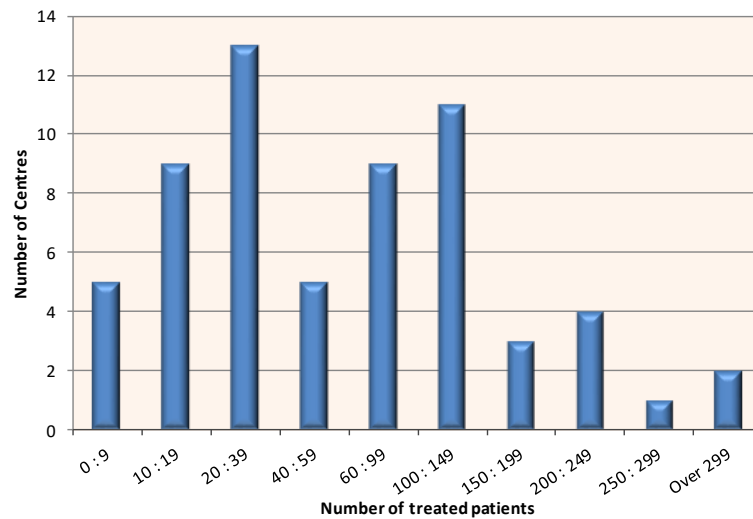
**Figure 25** Cumulative incidence chart of deaths from hepatocellular carcinoma or liver failure in UK patients with bleeding disorders 1969 - 2015



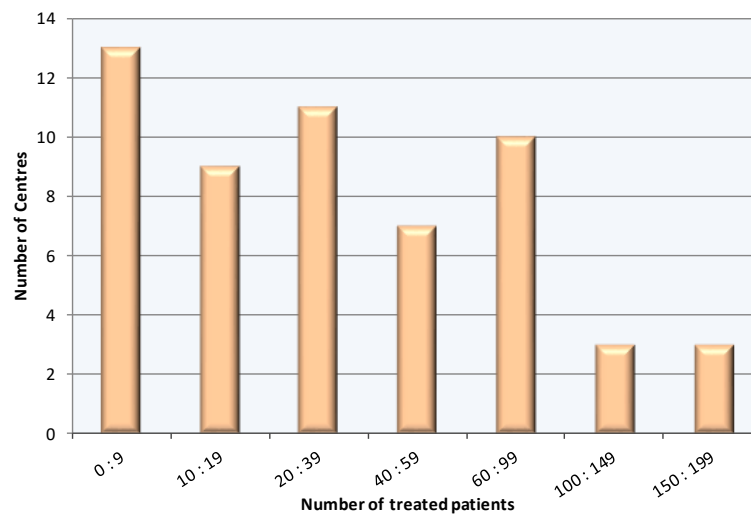
Year	Hepatocellular Carcinoma	Liver Failure	Total
1969	0	2	2
1970	0	2	2
1972	0	2	2
1973	0	1	1
1974	0	1	1
1975	0	1	1
1979	0	2	2
1980	0	1	1
1983	0	2	2
1984	0	3	3
1985	0	4	4
1986	0	7	7
1987	0	5	5
1988	1	2	3
1989	0	5	5
1990	0	4	4
1991	0	9	9
1992	0	9	9
1993	0	15	15
1994	0	13	13

Year	Hepatocellular Carcinoma	Liver Failure	Total
1995	0	7	7
1996	0	10	10
1997	0	10	10
1998	0	9	9
1999	0	4	4
2000	0	15	15
2001	1	6	7
2002	3	9	12
2003	2	2	4
2004	3	4	7
2005	1	2	3
2006	4	5	9
2007	2	3	5
2008	3	6	9
2009	2	8	10
2010	2	9	11
2011	2	5	7
2012	3	8	11
2013	9	9	18
2014	0	4	4
2015	2	9	11
<b>Total</b>	<b>40</b>	<b>232</b>	<b>272</b>

**Figure 26** Total number of patients with Haemophilia A, Haemophilia B or von Willebrand Disease treated by UK Haemophilia Centres



**Figure 27** Total number of patients with severe Haemophilia A and Haemophilia B treated by UK Haemophilia Centres



## Appendix 2

## Quarterly Returns - Participating Centres

Centre Name	
Aberdeen	Leicester
Abergavenny	Lewisham
Bangor	Lincoln
Barnstaple	Liverpool (R. I.)
Belfast - Adult's	Liverpool Children's
Belfast - Children's	Manchester (Adults)
Birmingham (Queen Elizabeth)	Manchester Children's
Birmingham Children's	Newcastle upon Tyne
Bournemouth / Poole	North Hampshire (Basingstoke)
Bradford	North Staffordshire (Stoke on Trent)
Brighton	Norwich
Bristol (Infirmary & Children's)	Nottingham
Cambridge	Oxford
Canterbury	Peterborough
Cardiff	Plymouth
Chichester	Portsmouth
Coventry	Royal Free
Derby	Salisbury
Dundee	Sheffield (Children's)
Edinburgh	Sheffield (Royal Hallamshire)
Exeter	Shrewsbury
Glasgow (R.H.S.C.)	Southampton
Glasgow (R.I.)	St George's Hospital, London
Great Ormond Street	St Thomas' and Guy's Hospital
Hammersmith Hospital, London	Swansea
Inverness	Taunton / Yeovil
Ipswich	The Royal London Hospital
Kettering	Torquay
Kingston upon Hull (Hull)	Truro
Lancaster	Wolverhampton
Leeds	York