

# UKHCDO Bleeding Disorder Statistics for April 2009 to March 2010

A report from the National Haemophilia Database

**UKHCDO**

UNITED KINGDOM HAEMOPHILIA CENTRES DOCTORS' ORGANISATION



**New Registrations - Number of patients newly registered at UK Haemophilia Centres between April 2009 & March 2010 showing their coagulation defect and gender**

Coagulation Defect	Male	Female	Total
Haemophilia A	168	-	168
Haemophilia B	29	-	29
Females with VIII deficiency	-	47	47
Females with IX deficiency	-	17	17
von Willebrand disease	135	271	406
F.V deficiency	1	3	4
F.VII deficiency	22	26	48
FIX Leyden	1	-	1
F.X deficiency	3	3	6
F.XI Deficiency	49	59	108
F.XIII Deficiency	4	1	5
Fibrinogen Deficiency	2	3	5
Prothrombin Deficiency	-	1	1
Dysfibrinogenaemia	3	11	14
Combined V+VIII Deficiency	-	1	1
Other combined diagnoses	14	11	25
Acquired Haemophilia A	31	34	65
Acquired von Willebrands	4	2	6
Acquired Deficiency (other)	1	-	1
Severe Platelet Disorders - Glanzmanns Thrombasthenia	2	2	4
Severe Platelet Disorders - Bernard Soulier	2	-	2
Other platelet defects	27	72	99
Miscellaneous	2	12	14
Unclassified	-	11	11
Pseudo vW	1	-	1
Probable von Willebrand's disease	5	11	16
<b>Total</b>	<b>506</b>	<b>598</b>	<b>1104</b>

**New Registrations of Haemophilia A & B between April 2009 & March 2010, by age at the end of March 2010 and disease severity**

Coagulation Defect	Age (years)	Number of Patients - Factor VIII/IX level iu/dl				
		≤1	>1 and <5	≥5	Not Known	Total
Haemophilia A	0 : 9	35	6	47	-	88
	10 : 19	5	-	21	-	26
	20 : 29	5	2	7	-	14
	30 : 39	3	3	9	-	15
	40 : 49	1	-	8	-	9
	50 : 59	-	-	5	-	5
	60 : 69	0	2	3	-	5
	70 : 79	0	-	3	-	3
	80 : 89	0	-	3	-	3
<b>Total</b>		<b>49</b>	<b>13</b>	<b>106</b>	<b>0</b>	<b>168</b>
Haemophilia B	0 : 9	8	3	4	-	15
	10 : 19	1	-	3	-	4
	20 : 29	-	4	1	-	5
	30 : 39	-	-	2	1	3
	40 : 49	-	-	1	-	1
	50 : 59	-	-	1	-	1
<b>Total</b>		<b>9</b>	<b>7</b>	<b>12</b>	<b>1</b>	<b>29</b>

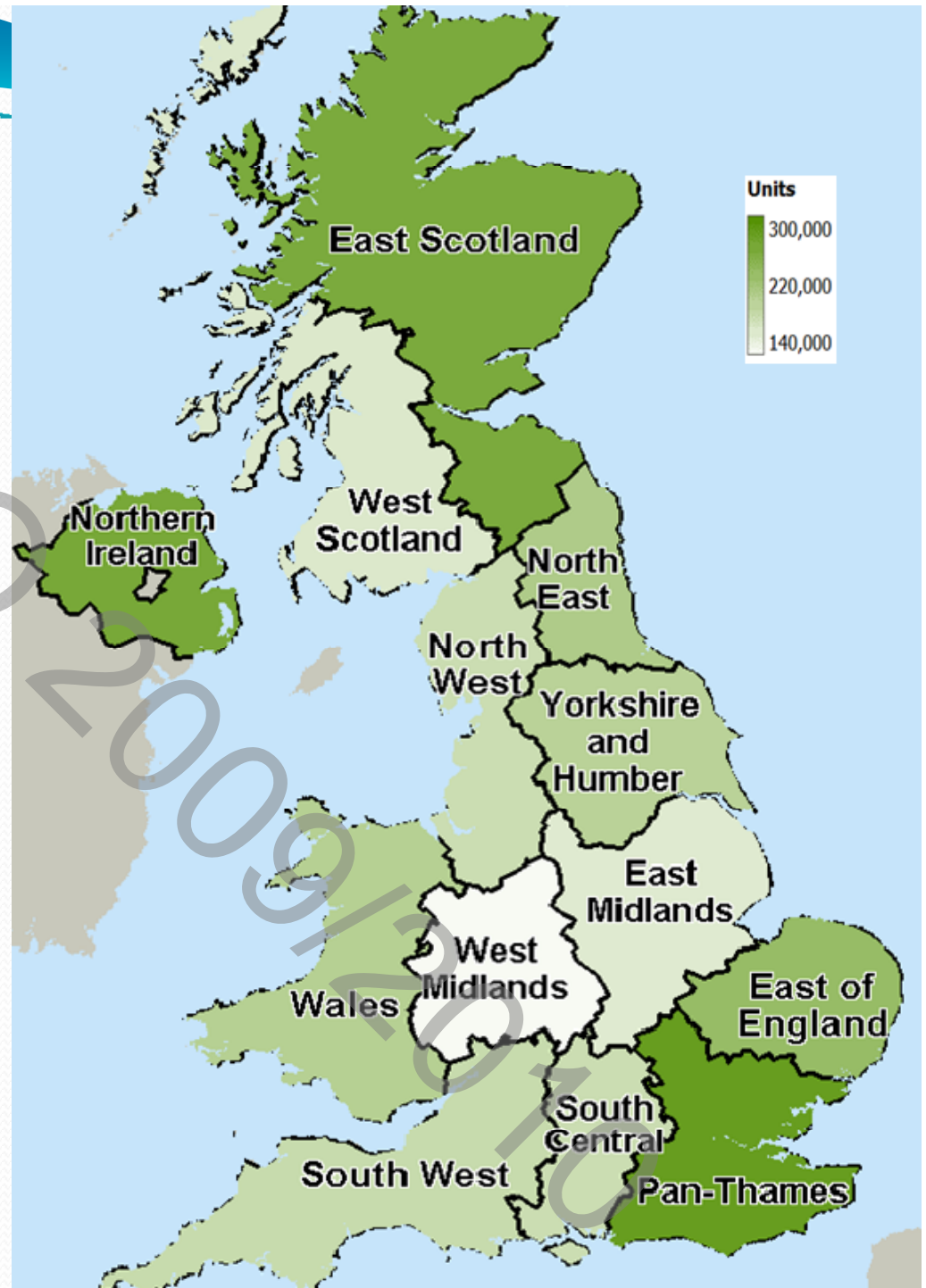
**In Register – The total Number of patients with all types of bleeding disorder currently in the register and the number treated during April 2009 & March 2010**

Coagulation Defect	Number of Patients		Percent Treated
	In Register	Treated	
Haemophilia A	5,346	2,863	53.55%
Haemophilia B	1,125	618	54.93%
Females with VIII deficiency	1,082	53	4.90%
Females with IX deficiency	332	37	11.14%
von Willebrand disease	8,879	983	11.07%
F.V deficiency	130	8	6.15%
F.VII deficiency	641	47	7.33%
FIX Leyden	8	0	0.00%
FIX Leyden Carrier	1	1	100.00%
F.X deficiency	180	32	17.78%
F.XI Deficiency	1,833	75	4.09%
F.XIII Deficiency	56	46	82.14%
Fibrinogen Deficiency	172	17	9.88%
Prothrombin Deficiency	9	1	11.11%
Dysfibrinogenaemia	144	6	4.17%
Combined II+VII+IX+X Deficiency	4	0	0.00%
Combined V+VIII Deficiency	23	6	26.09%
Other combined diagnoses	209	25	11.96%
Acquired Haemophilia A	309	63	20.39%
Acquired Haemophilia B	2	0	0.00%
Acquired von Willebrands	68	14	20.59%
Acquired F.XIII Deficiency	1	0	0.00%
Acquired Deficiency (other)	3	2	66.67%
Severe Platelet Disorders - Glanzmann's Thrombasthenia	103	28	27.18%
Severe Platelet Disorders - Bernard Soulier	57	8	14.04%
Other platelet defects	1,219	57	4.68%
Haemophilia A with Liver Transplant	12	1	8.33%
Haemophilia B with Liver Transplant	5	0	0.00%
Pseudo vW	1	0	0.00%
Probable von Willebrand's disease	31	7	22.58%
Miscellaneous	104	6	5.77%
Unclassified	100	5	5.00%
<b>Total</b>	<b>22,189</b>	<b>5,009</b>	<b>22.57%</b>

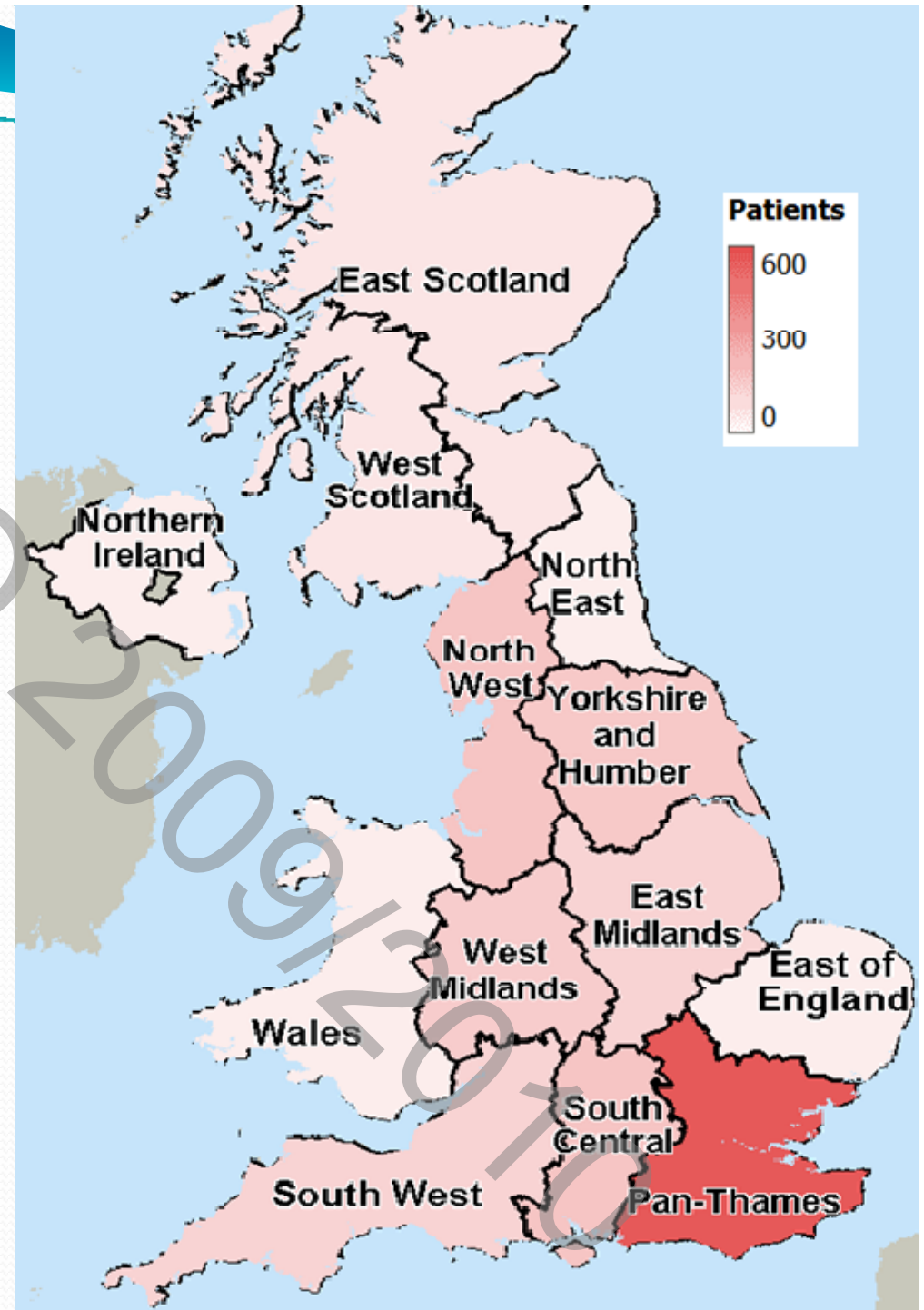
**In Register – The total number of haemophilia A, B and von Willebrand's disease patients currently in the register, by severity, age group and gender**

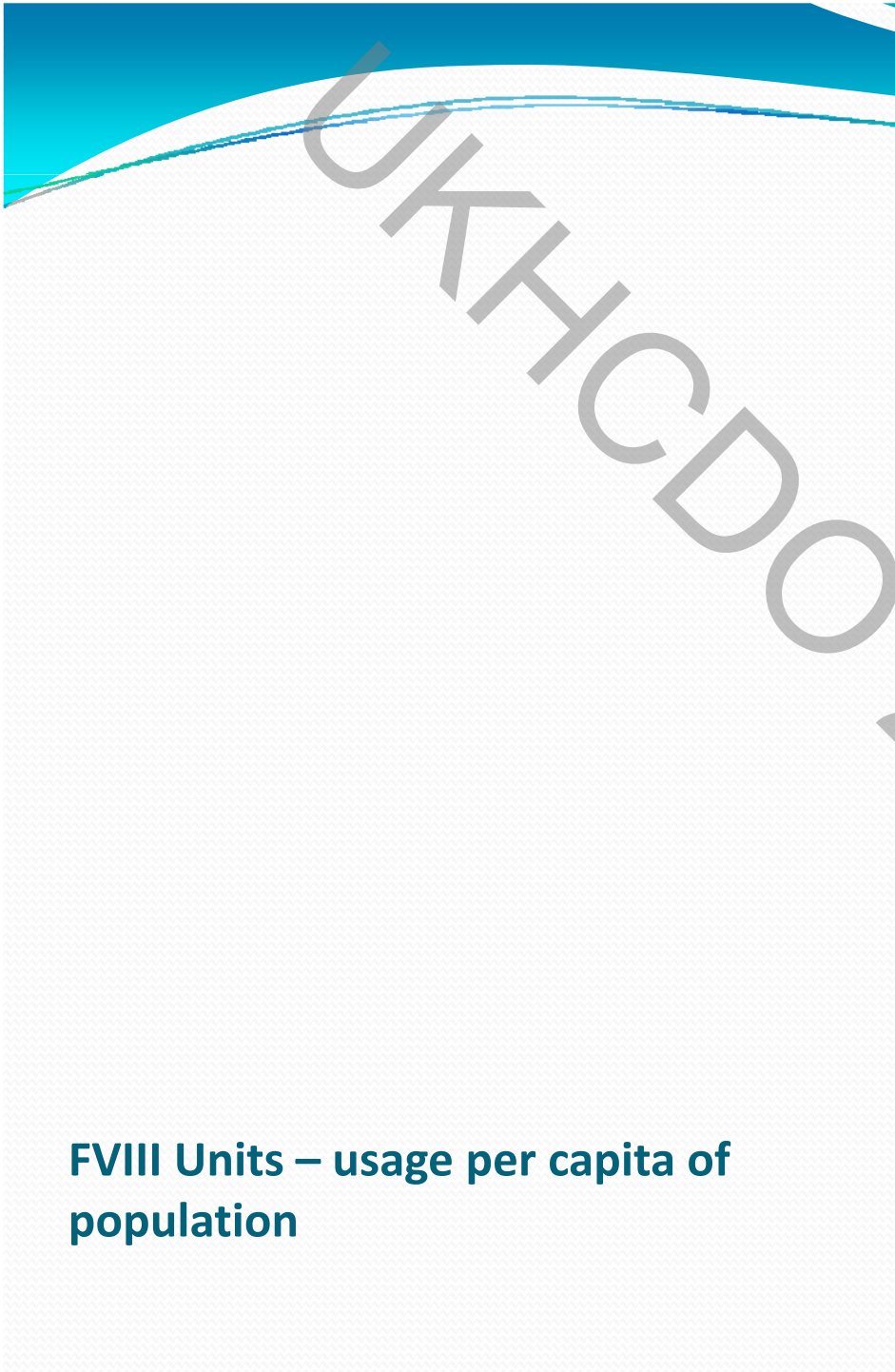
Coagulation Defect	Males										Females										Overall Total
	<18 years					=>18 years					<18 years					=>18 years					
	≤ 1	>1 & <5	≥ 5	N/K	Sub Total	≤ 1	>1 & <5	≥ 5	N/K	Sub Total	≤ 1	>1 & <5	≥ 5	N/K	Sub Total	≤ 1	>1 & <5	≥ 5	N/K	Sub Total	
Haemophilia A	645	154	654	0	<b>1453</b>	1155	401	2299	1	<b>3856</b>	-	1	-	-	<b>1</b>	-	-	-	-	<b>0</b>	<b>5310</b>
Haemophilia B	117	58	92	0	<b>267</b>	274	183	384	3	<b>844</b>	-	-	-	-	<b>0</b>	-	1	-	-	<b>1</b>	<b>1112</b>
Female with FVIII deficiency	-	-	-	-	-	-	-	-	-	-	5	8	96	12	<b>121</b>	32	15	874	32	<b>953</b>	<b>1074</b>
Female with FIX deficiency	-	-	-	-	-	-	-	-	-	-	3	2	48	9	<b>62</b>	6	7	245	9	<b>267</b>	<b>329</b>
von Willebrand disease	27	23	841	24	<b>915</b>	37	45	2232	39	<b>2353</b>	11	28	785	31	<b>855</b>	50	68	4505	110	<b>4733</b>	<b>8856</b>

**Factor VIII Usage (inc. inhibitor)  
divided by number of Severe  
Haemophilia A patients by  
commissioning region**

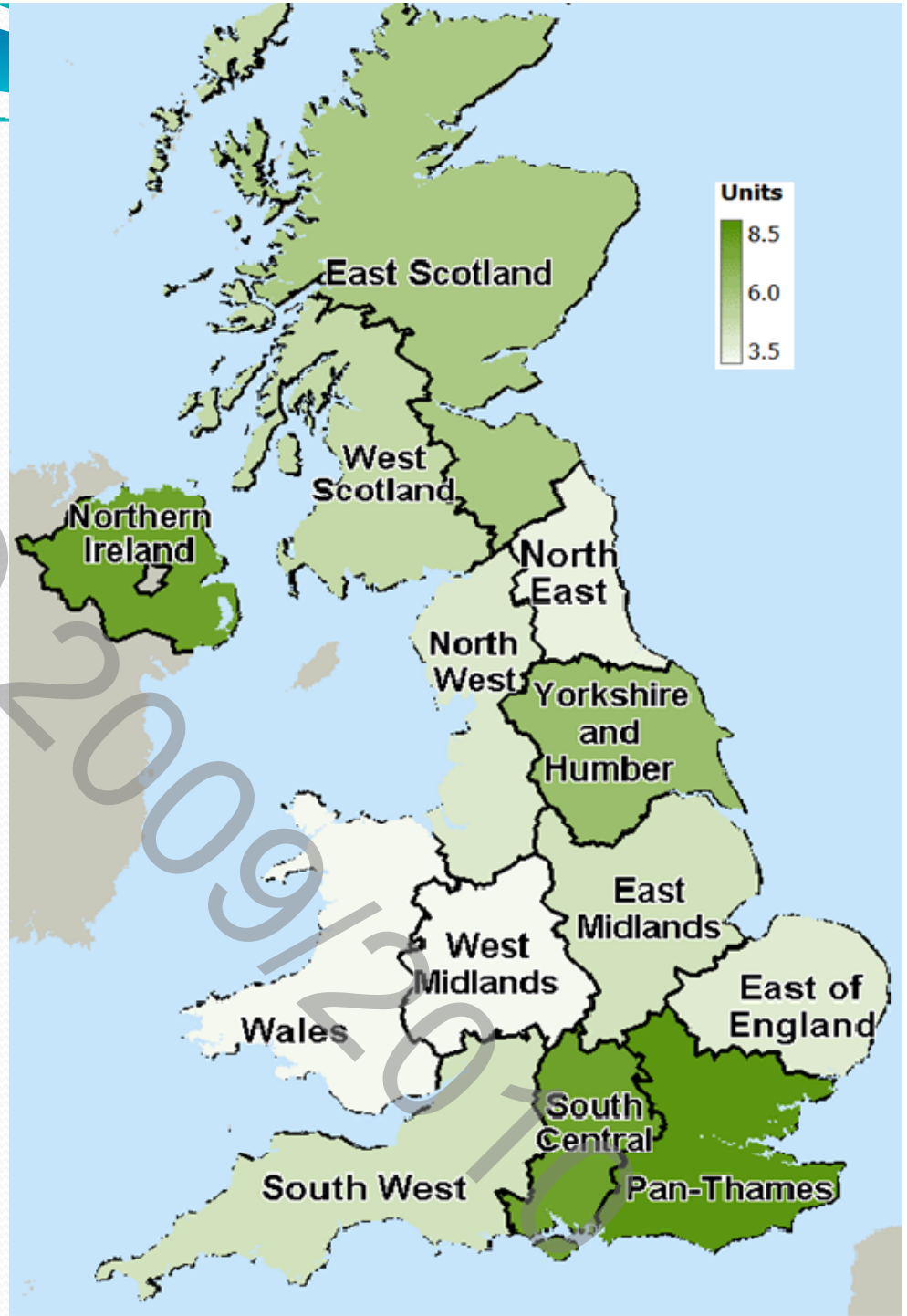


**Total Number of Severe Haemophilia A & B Patients by commissioning region**

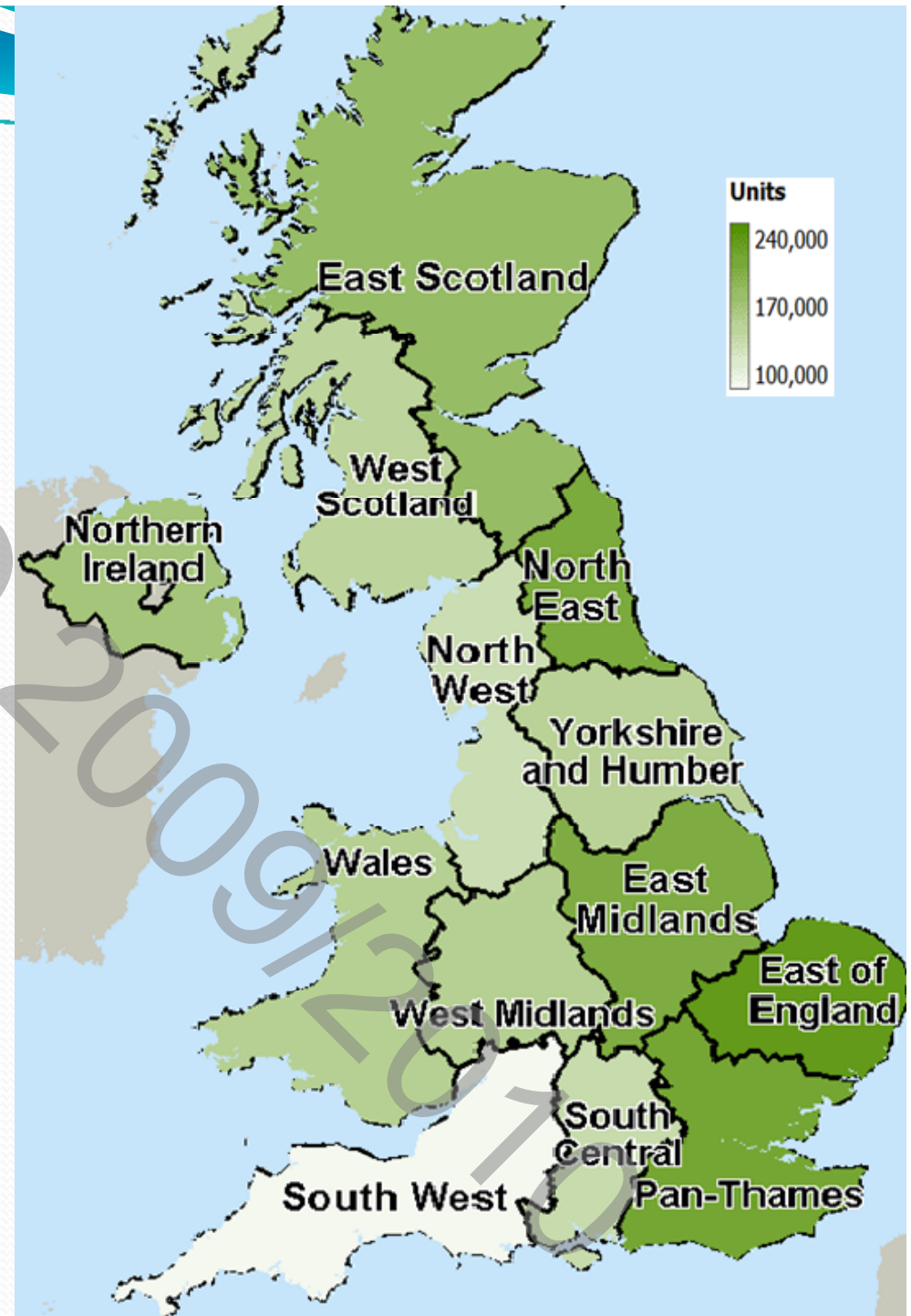




**FVIII Units – usage per capita of population**



Factor IX Usage (inc. inhibitor)  
divided by number of Severe  
Haemophilia B patients by  
commissioning region



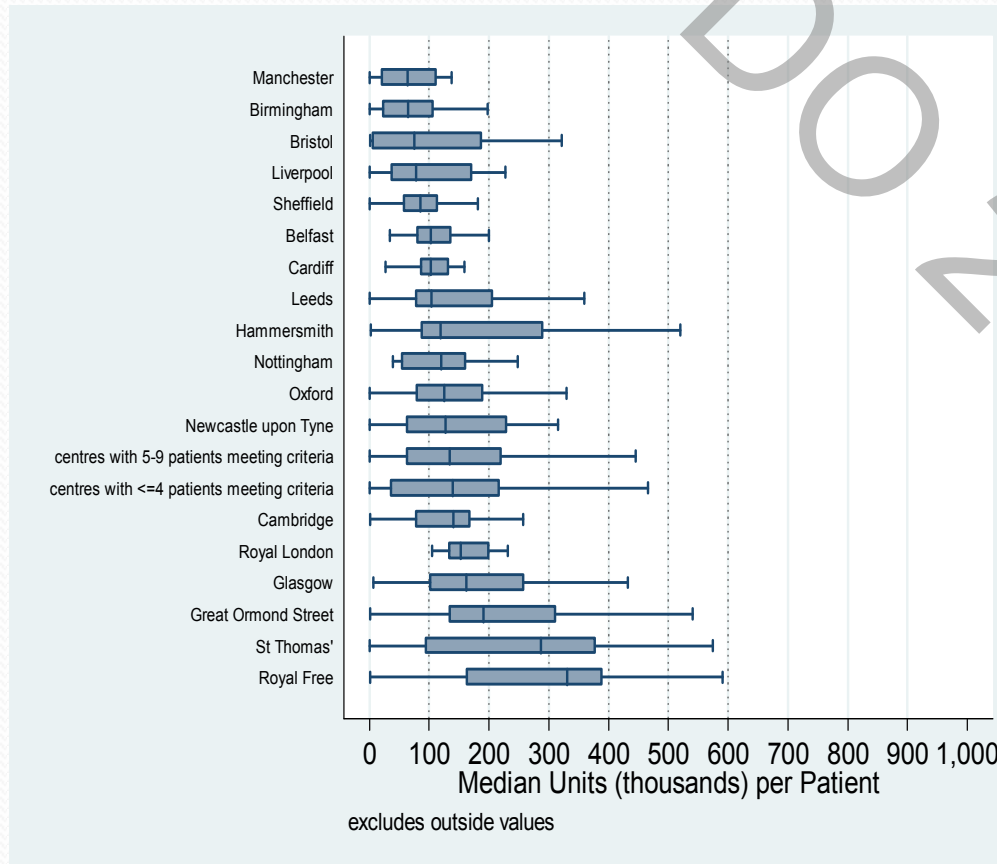
## Factor VIII Usage by commissioning region for Severe Haemophilia A patients (inc. inhibitor)

SCG / Region	Population *	Number of Severe Haemophilia A Patients	Total FVIII Units Used	Mean Usage Haemophilia A	FVIII Units Per Capita
Pan-Thames Haemophilia Consortium	15,318,900	432	127,142,342	294,311	8.30
East Midlands Specialised Commissioning Group	4,433,000	123	21,277,810	172,990	4.80
East of England Specialised Commissioning Group	2,335,500	41	10,130,700	247,090	4.34
North East Specialised Commissioning Group	2,575,500	49	10,679,340	217,946	4.15
North West Specialised Commissioning Group	6,875,700	143	25,279,000	176,776	3.68
South West Specialised Commissioning Group	5,209,200	120	25,330,516	211,088	4.86
South Central Specialised Commissioning Group	4,062,300	149	31,436,476	210,983	7.74
West Midlands Specialised Commissioning Group	5,411,100	133	20,728,959	155,857	3.83
Yorkshire and Humber Specialised Commissioning Group	5,213,200	155	33,565,808	216,554	6.44
Wales	2,993,400	68	17,669,732	259,849	5.90
East Scotland	2,953,429	64	17,513,013	273,641	5.93
West Scotland	2,215,071	67	11,639,899	173,730	5.25
Northern Ireland	1,775,000	52	13,882,576	266,973	7.82

## Factor IX Usage by commissioning region for Severe Haemophilia B patients (inc. inhibitor)

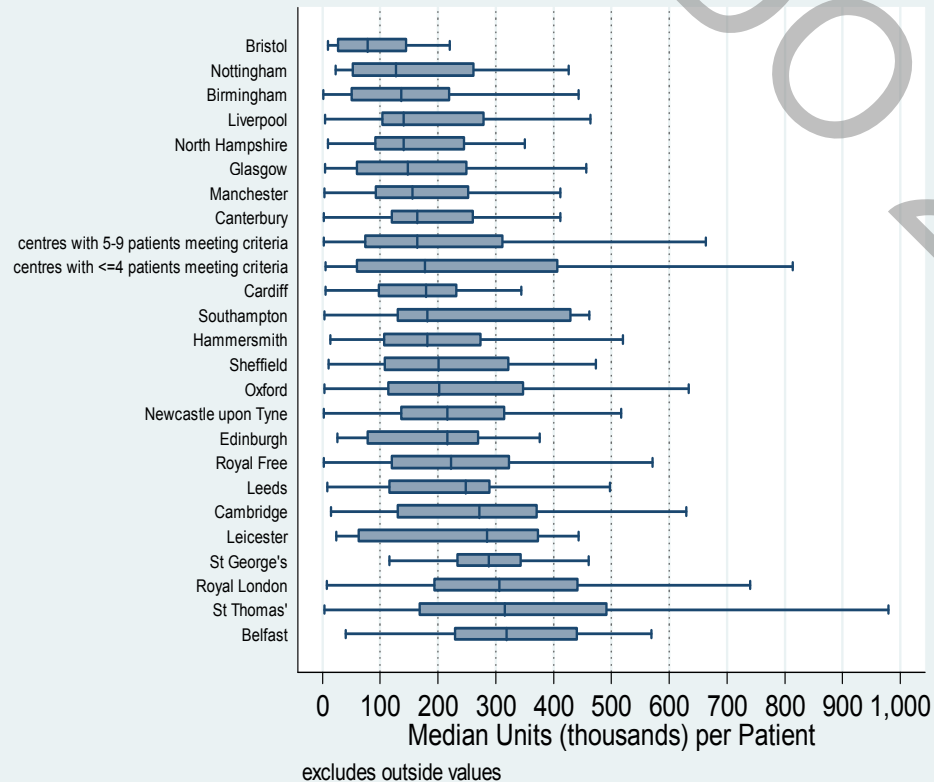
SCG / Region	Population *	Number of Severe Haemophilia B Patients	Total FIX Units Used	Mean Usage Haemophilia B	FIX Units Per Capita
Pan-Thames Haemophilia Consortium	15,318,900	112	24,604,639	219,684	1.61
East Midlands Specialised Commissioning Group	4,433,000	20	4,112,181	205,609	0.93
East of England Specialised Commissioning Group	2,335,500	18	4,101,570	227,865	1.76
North East Specialised Commissioning Group	2,575,500	12	2,493,200	207,767	0.97
North West Specialised Commissioning Group	6,875,700	28	4,311,845	153,994	0.63
South West Specialised Commissioning Group	5,209,200	15	1,855,940	123,729	0.36
South Central Specialised Commissioning Group	4,062,300	23	3,806,840	165,515	0.94
West Midlands Specialised Commissioning Group	5,411,100	27	4,516,674	167,284	0.83
Yorkshire and Humber Specialised Commissioning Group	5,213,200	24	3,897,979	162,416	0.75
Wales	2,993,400	12	1,923,000	160,250	0.64
East Scotland	2,953,429	12	2,420,608	201,717	0.82
West Scotland	2,215,071	21	3,409,073	162,337	1.54
Northern Ireland	1,775,000	5	1,041,000	208,200	0.59

## FVIII usage in severe haemophilia A patients with no current inhibitor by centre, aged less than 18 years



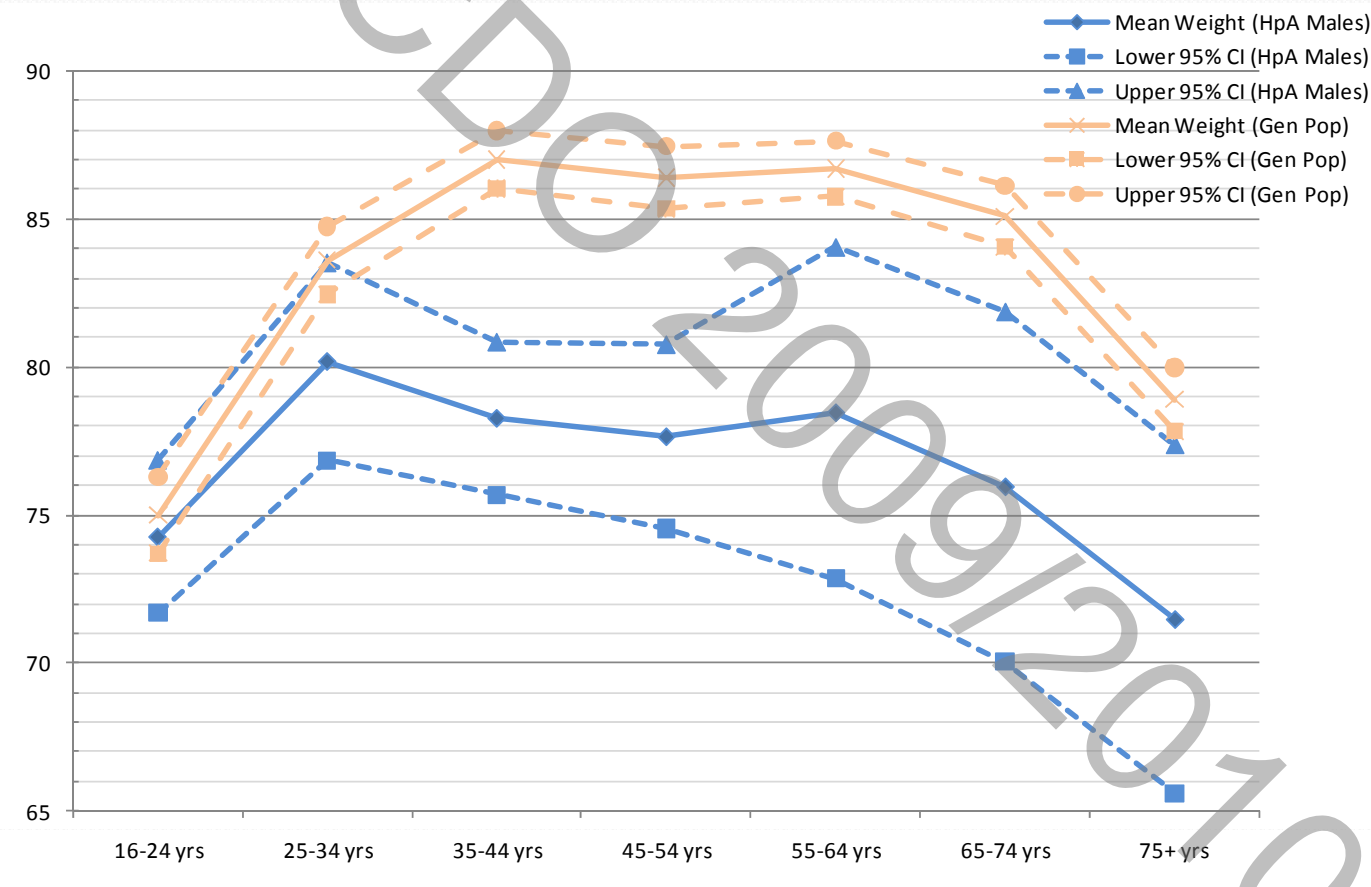
Centre	Patients	Total Units	Median Units
Belfast	21	2,355,076	103,000
Birmingham	42	3,350,750	64,875
Bristol	12	1,537,750	75,500
Cambridge	19	2,595,500	140,000
Cardiff	12	1,402,250	103,000
Glasgow	26	4,967,750	162,375
Great Ormond Street	60	13,421,500	191,000
Hammersmith	14	2,519,000	119,000
Leeds	28	4,074,500	103,750
Liverpool	19	1,807,000	78,250
Manchester	20	1,314,250	63,750
Newcastle upon Tyne	25	3,891,000	127,000
Nottingham	11	1,336,875	119,918
Oxford	41	5,601,900	125,000
Royal Free	26	7,486,160	330,500
Royal London	13	1,993,640	153,250
Sheffield	29	2,713,000	84,950
St Thomas'	30	7,646,500	287,000
centres with <=4 patients meeting criteria	41	6,156,934	139,078
centres with 5-9 patients meeting criteria	86	13,010,276	134,500

## FVIII usage in severe haemophilia A patients with no current inhibitor by centre, aged 18 years or more

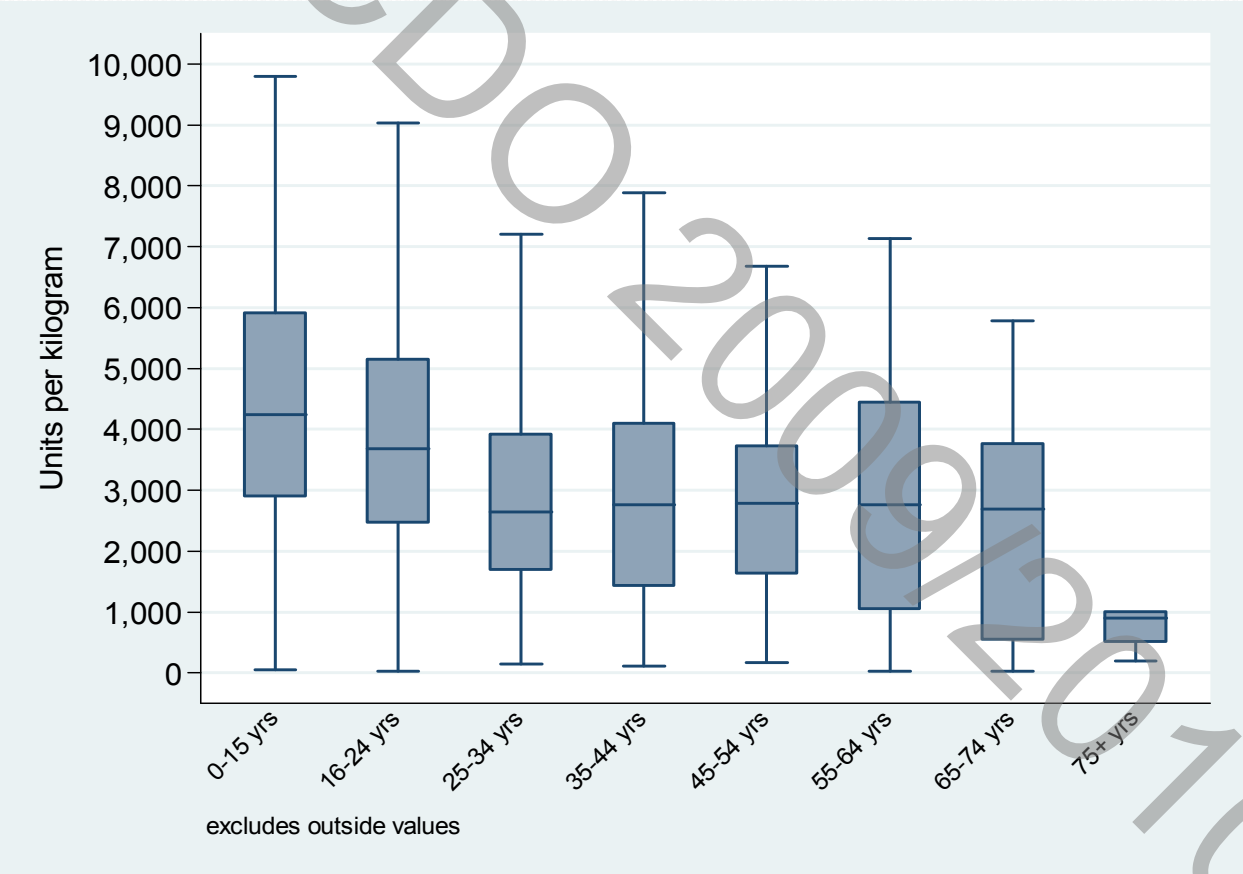


Centre	Patients	Total Units	Median Units
Bristol	18	1,634,500	78,250
Nottingham	27	4,571,617	127,000
Birmingham	68	10,774,000	136,250
Liverpool	29	5,558,000	140,000
North Hampshire	32	5,409,000	140,500
Glasgow	42	7,169,821	148,112
Manchester	69	11,853,250	155,500
Canterbury	18	3,613,500	164,000
centres with 5-9 patients meeting criteria	124	26,172,689	164,250
centres with <=4 patients meeting criteria	27	6,902,000	177,000
Cardiff	26	5,179,750	179,250
Hammersmith	15	3,195,500	181,500
Southampton	14	3,227,500	181,500
Sheffield	38	8,349,100	201,000
Oxford	84	19,177,400	202,000
Newcastle upon Tyne	33	7,966,000	216,000
Edinburgh	19	4,970,829	216,211
Royal Free	86	19,927,070	222,750
Leeds	25	6,475,250	248,000
Cambridge	19	5,337,700	271,500
Leicester	15	3,476,000	285,000
St George's	22	6,492,500	287,500
Royal London	35	11,488,680	306,000
St Thomas'	98	32,836,000	315,000
Belfast	29	9,610,500	319,000

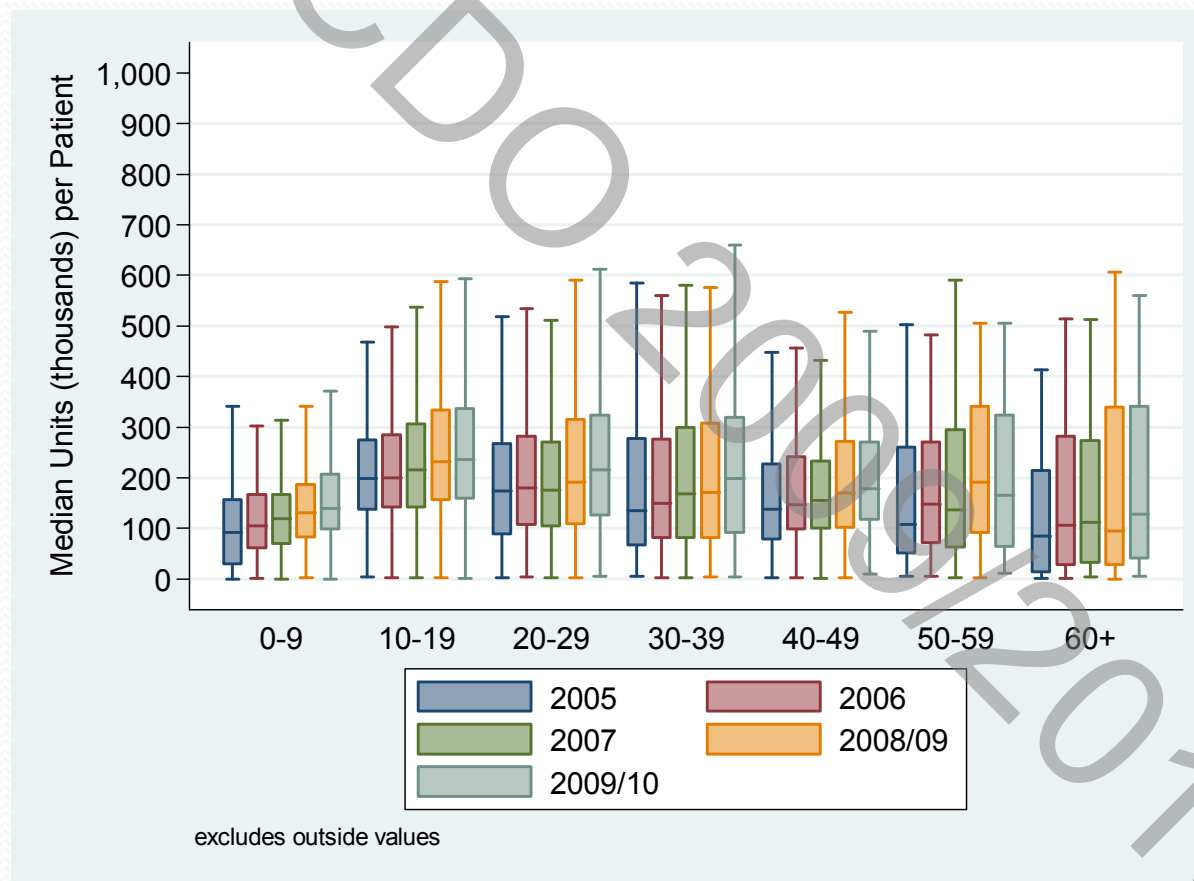
# Mean weight of Severe Haemophilia A males compared to the general population



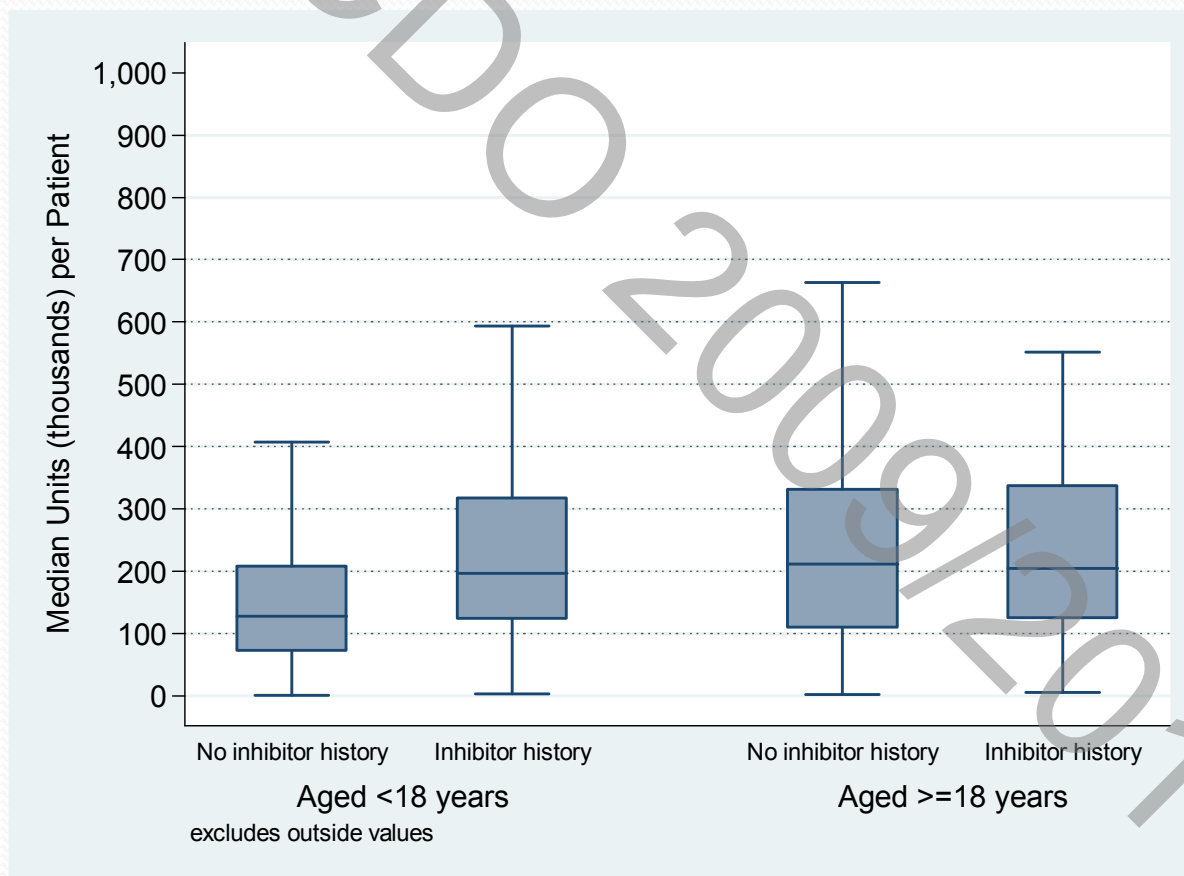
# Median FVIII units used per kilogram body weight per year in Severe Haemophilia A patients, 2009/10



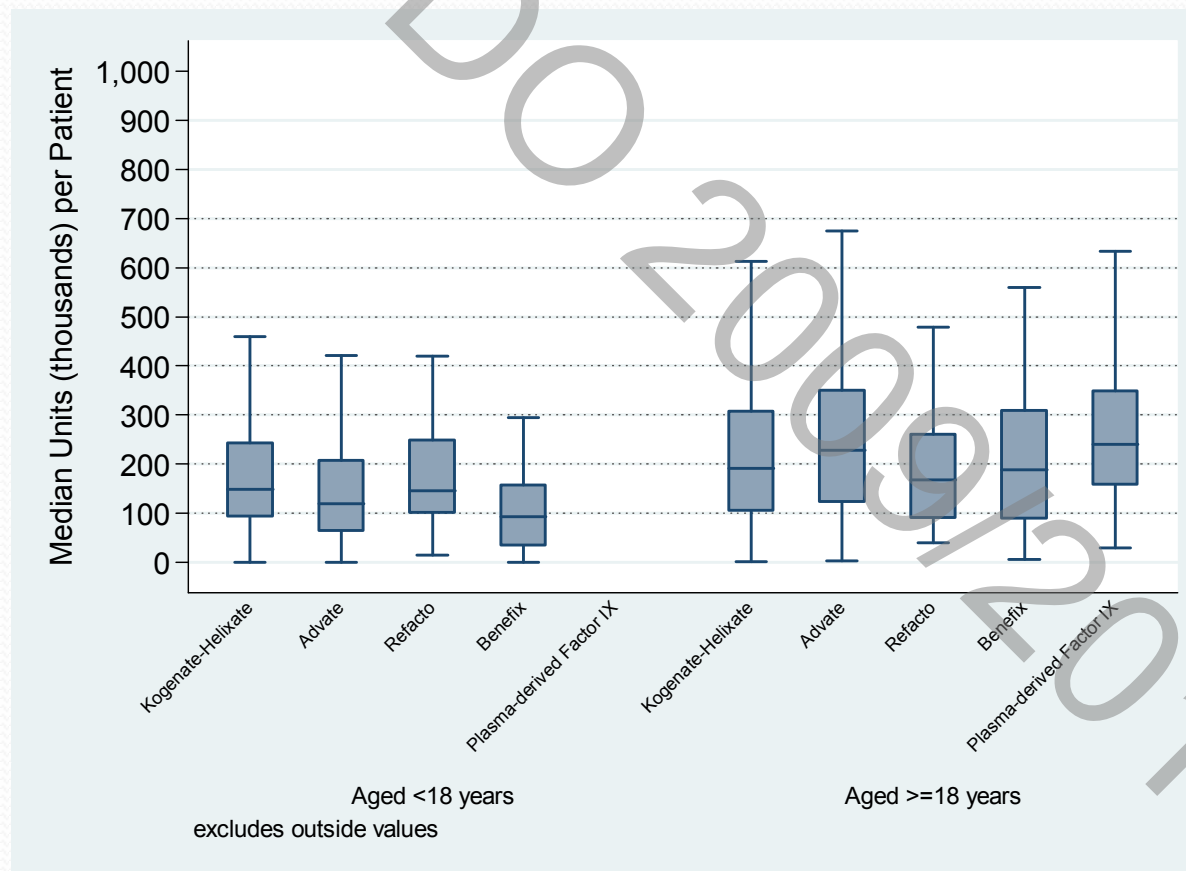
## Severe Haemophilia A patients treated with FVIII products with no current inhibitor: median IU issued



## Severe Haemophilia A patients with no current inhibitor between April 2009 & March 2010: median usage by inhibitor history



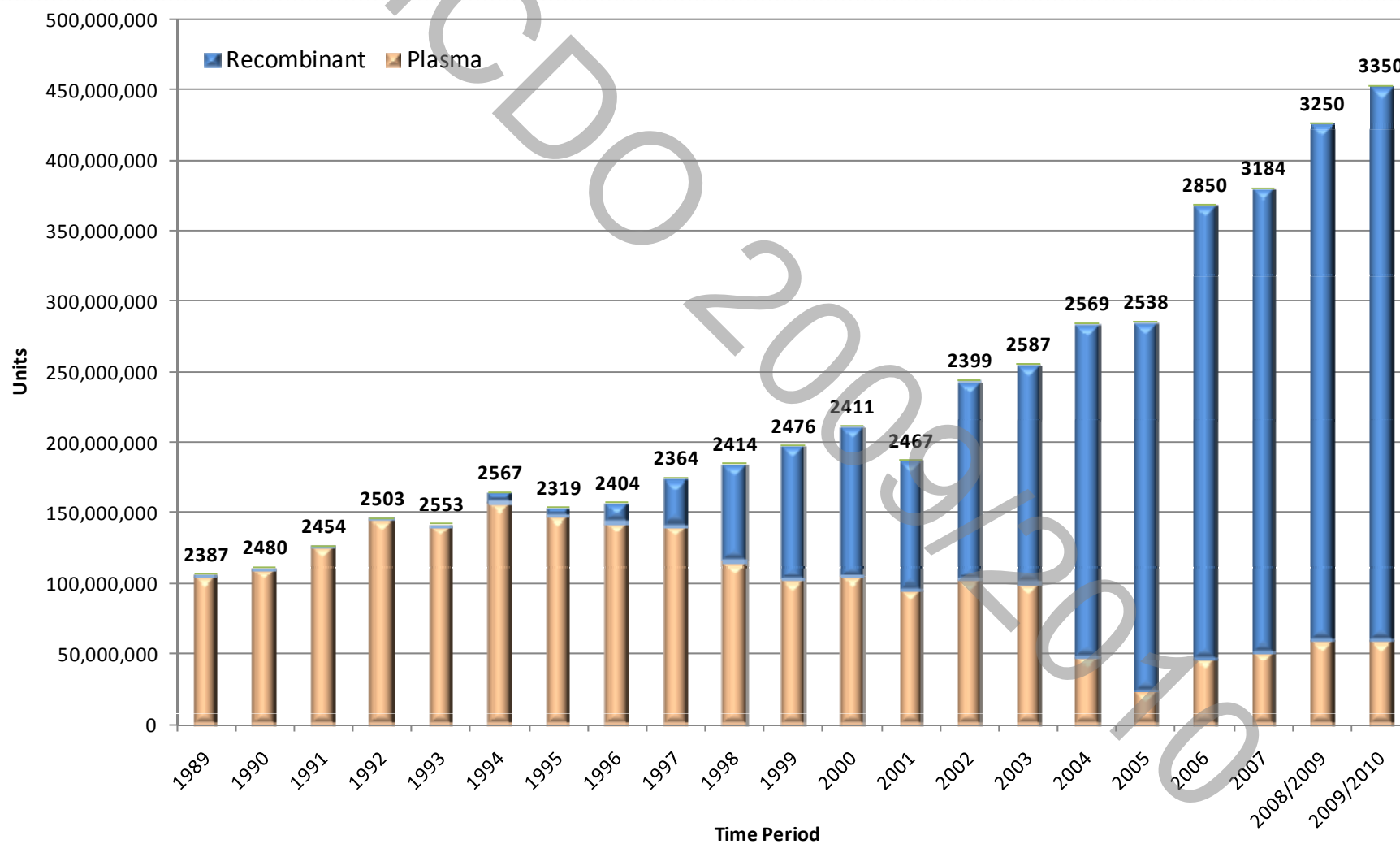
## Severe Haemophilia A & B patients with no current inhibitor using only one product between April 2009 & March 2010: median usage



## Factor VIII units used by UK Haemophilia Centres

Coagulation Defect	Patients Treated	Plasma FVIII	Recombinant FVIII	Total FVIII
Haemophilia A	2,669	40,041,800	390,375,395	430,417,195
Females with VIII deficiency	31		457,011	457,011
von Willebrand disease	585	17,390,135	46,082	17,436,217
Combined V+VIII Deficiency	6		29,598	29,598
Other combined diagnoses	15	1,759,500	1,017,250	2,776,750
Acquired Haemophilia A	7		214,744	214,744
Acquired von Willebrands	13	575,500	5,000	580,500
Miscellaneous	3	38,500	7,500	46,000
<b>Total</b>	<b>3,329</b>	<b>59,805,435</b>	<b>392,152,580</b>	<b>451,958,015</b>

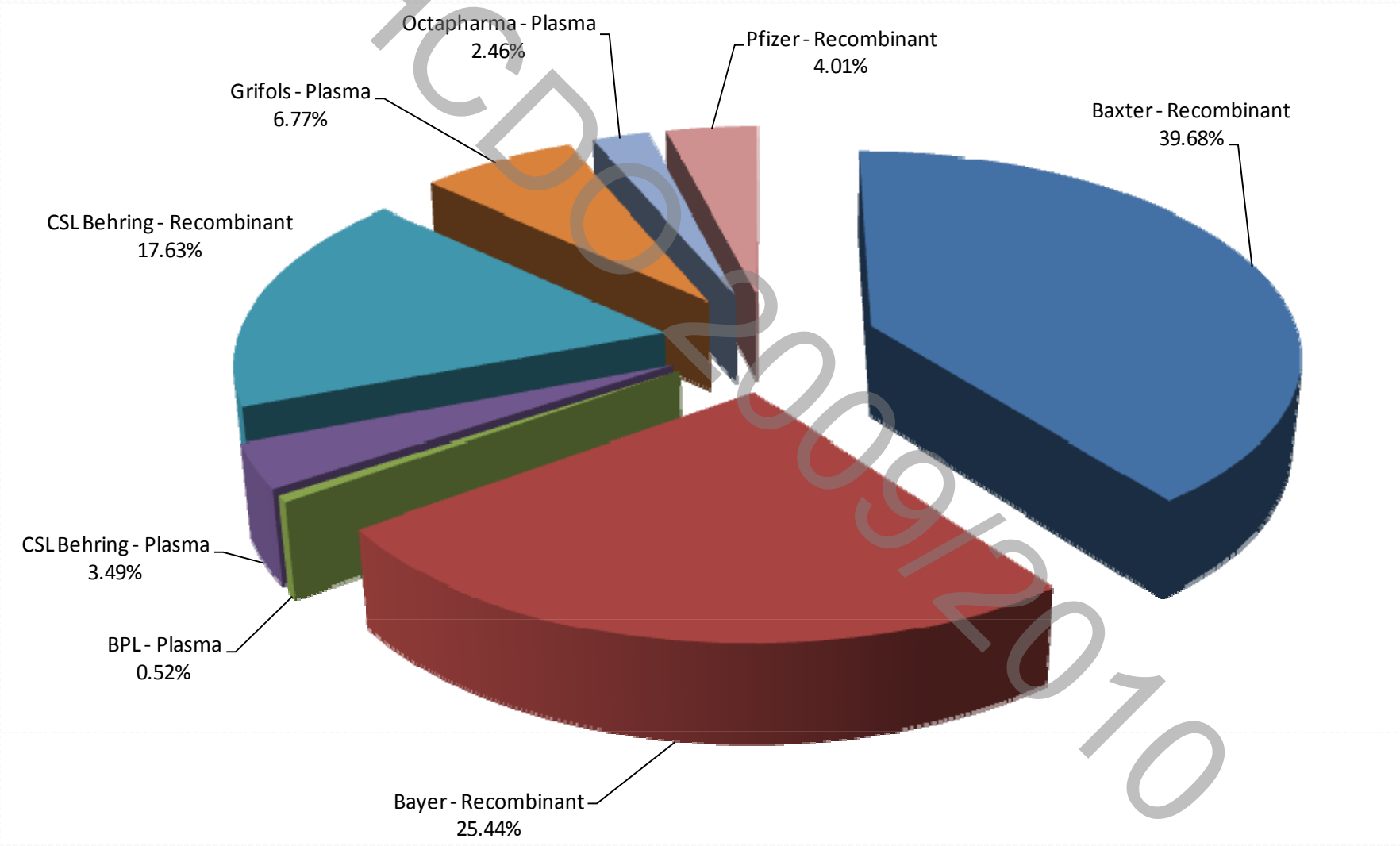
## Factor VIII units by UK Haemophilia Centres: by calendar year between 1989 and 2007 and by financial year from April 2008 onwards



**Products used for  
Haemophilia A (including  
inhibitors)**

Manufacturer	Product	Total Units
	Desmopressin-Octim Sub-cut (15mcg/ml)	5,645
	Desmopressin-DDAVP (4mcg/ml)	1,265
	Desmopressin-Octim nasal spray (150mcg/2.5ml vial)	64,090
	Haemocompletten P (g)	28
<b>Baxter</b>	Advate	179,032,922
	FEIBA	19,873,181
<b>Bayer</b>	Kogenate	114,536,110
<b>BPL</b>	FVIII 8Y	803,820
	Optivate	1,340,690
<b>CSL Behring</b>	Beriplex	4,000
	Haemate P	311,500
	Helixate Nexgen	78,804,294
<b>Grifols</b>	Alphanate	59,000
	Fanhdi	26,419,790
<b>Novo Nordisk</b>	NovoSeven (mg)	23,232.4
<b>Octapharma</b>	Octanate	11,107,000
	Octaplas	2
	Octaplex	1,000
<b>Pfizer</b>	ReFacto AF	6,618,765
	Refacto	11,381,804

# Market Share of factor VIII concentrates known to have been used by UK Haemophilia Centres

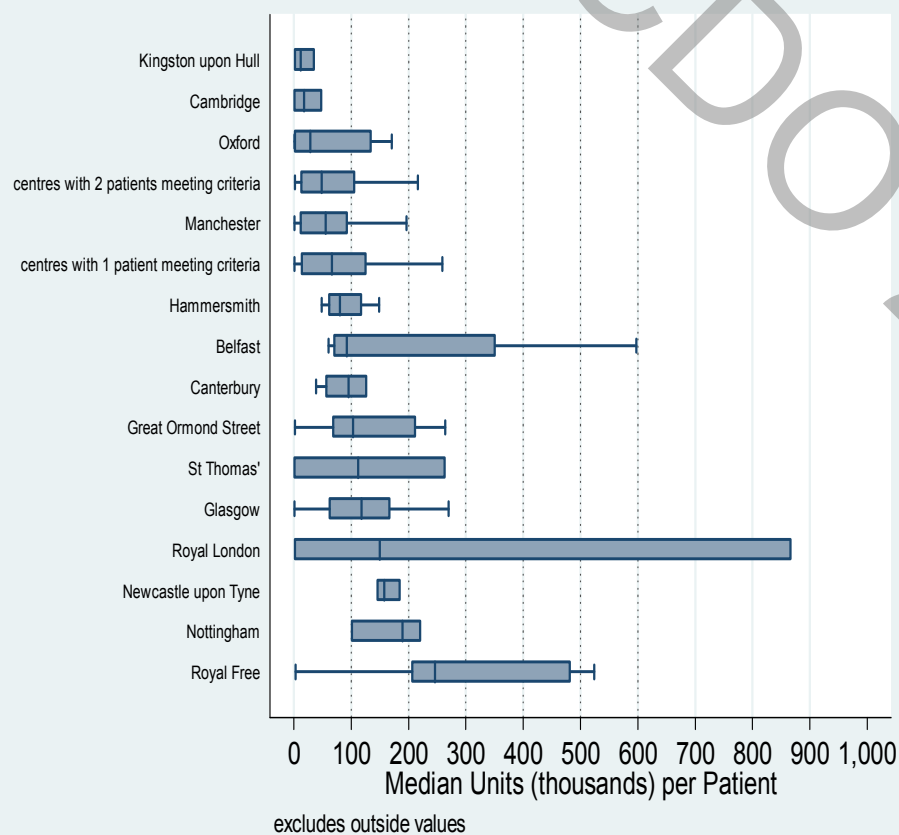


## Inhibitors by disease severity

Diagnosis	Number of Patients ever known to have an inhibitor by disease severity								
	≤ 1 iu/dl			>1 and <5 iu/dl			≥ 5 iu/dl		
	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%
Haemophilia A	1814	351	19.35%	559	43	7.69%	2972	62	2.09%
Haemophilia B	396	15	3.79%	244	0	0.00%	482	0	0.00%
von Willebrand disease	125	2	1.60%	164	2	1.22%	8381	6	0.07%

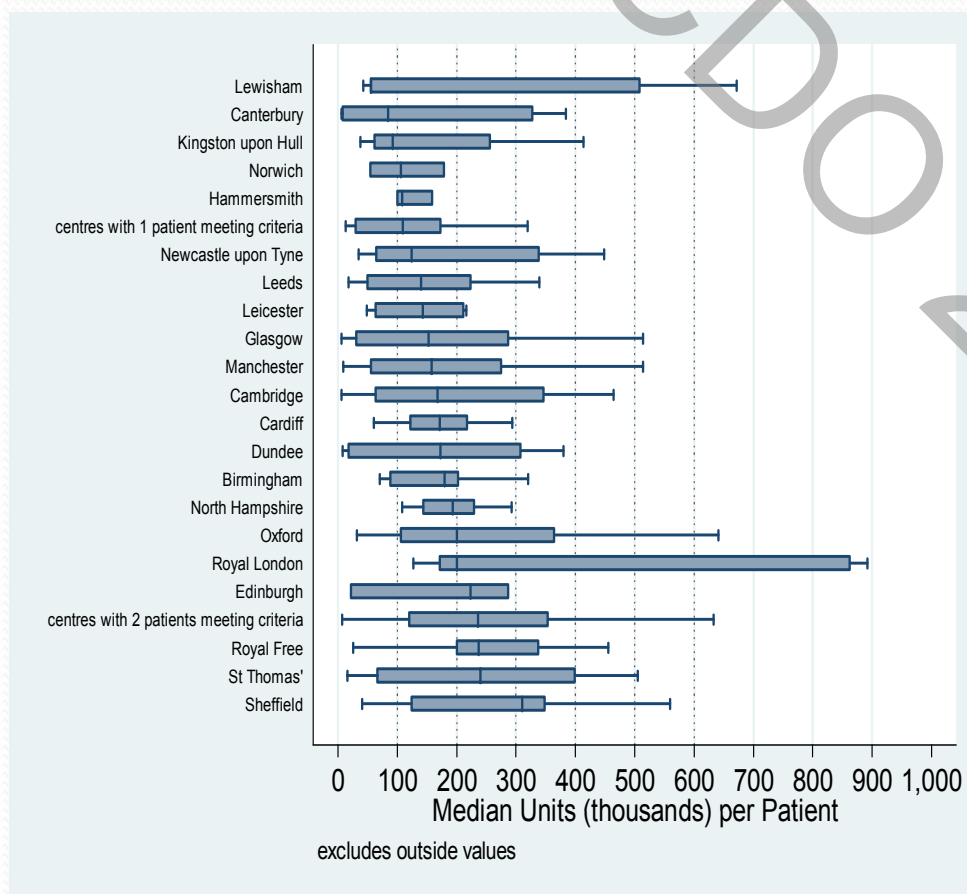
Diagnosis	Patients with a current inhibitor treated between April 2009 and March 2010 by disease severity								
	≤ 1 iu/dl			>1 and <5 iu/dl			≥ 5 iu/dl		
	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%
Haemophilia A	1655	146	8.82%	358	13	3.63%	848	20	2.36%
Haemophilia B	335	10	2.99%	134	0	0.00%	147	0	0.00%
von Willebrand disease	47	2	4.26%	50	1	2.00%	862	4	0.46%

## FIX usage in severe Haemophilia B patients with no current inhibitor by centre, aged less than 18 years



Centre	Patients	Total Units	Median Units
Belfast	4	842,000	92,500
Cambridge	3	67,500	18,000
Canterbury	6	815,000	95,000
Glasgow	5	619,000	117,750
Great Ormond Street	18	2,430,250	103,500
Hammersmith	4	358,010	80,505
Kingston upon Hull	3	49,140	11,838
Manchester	10	763,000	55,500
Newcastle upon Tyne	5	989,500	158,000
Nottingham	3	511,512	189,420
Oxford	10	630,500	29,000
Royal Free	5	1,461,530	246,390
Royal London	3	1,018,722	150,000
St Thomas'	3	377,000	112,000
centres with 1 patient meeting criteria	14	1,192,941	66,500
centres with 2 patients meeting criteria	16	1,065,250	48,250

## FIX usage in severe Haemophilia B patients with no current inhibitor by centre, aged 18 years or more

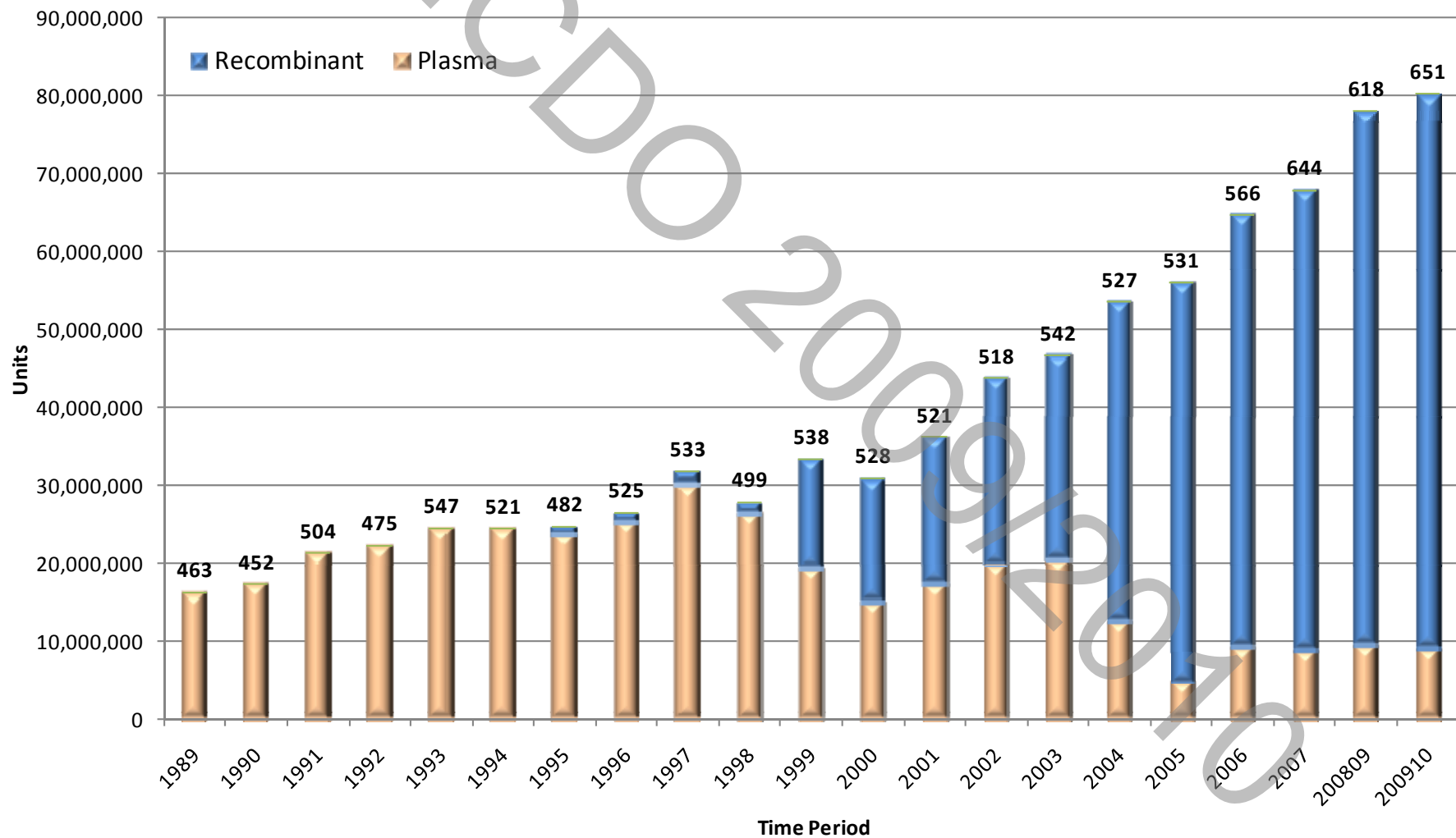


Centre	Patients	Total Units	Median Units
Birmingham	9	1,779,000	180,000
Cambridge	17	3,476,000	168,000
Canterbury	6	894,000	84,000
Cardiff	9	1,555,500	172,000
Dundee	6	1,058,983	172,790
Edinburgh	3	532,420	223,024
Glasgow	16	2,664,073	152,500
Hammersmith	3	368,000	108,000
Kingston upon Hull	4	635,729	91,903
Leeds	5	770,500	139,500
Leicester	4	550,960	142,980
Lewisham	5	1,335,426	55,808
Manchester	16	2,983,845	158,000
Newcastle upon Tyne	11	2,075,000	124,000
North Hampshire	6	1,161,000	193,000
Norwich	3	340,500	106,000
Oxford	21	5,359,880	200,000
Royal Free	25	6,369,598	237,224
Royal London	7	2,748,688	200,120
Sheffield	9	2,376,575	310,000
St Thomas'	15	3,534,750	240,000
centres with 1 patient meeting criteria	14	2,045,668	109,250
centres with 2 patients meeting criteria	14	4,215,099	236,000

## Factor IX units used by UK Haemophilia Centres

Coagulation Defect	Patients Treated	Plasma FIX	Recombinant FIX	Total FIX Units
Haemophilia B	611	9,137,700	70,438,519	79,576,219
Females with IX deficiency	37	-	478,738	478,738
FIX Leyden Carrier	1	-	14,000	14,000
Combined diagnoses	1	-	106,000	106,000
<b>Total</b>	<b>650</b>	<b>9,137,700</b>	<b>71,037,257</b>	<b>80,174,957</b>

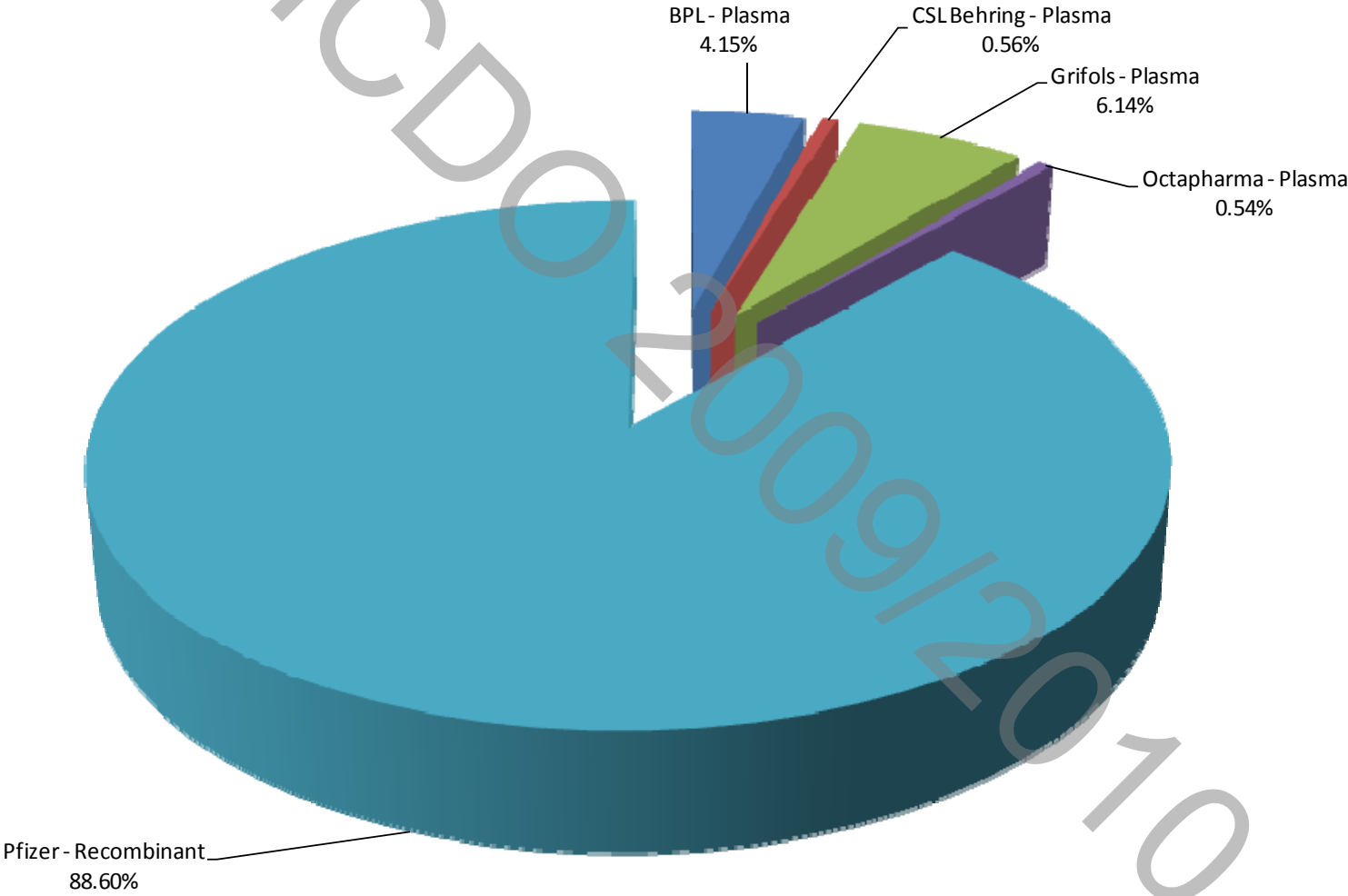
## Total factor IX units by UK Haemophilia Centres: by calendar year between 1989 and 2007 and by financial year from April 2008 onwards



## Products used for Haemophilia B (including inhibitors)

Manufacturer	Product	Total Units
Baxter	FEIBA	1,819,000
BPL	Replenine	3,326,370
CSL Behring	Mononine	451,000
Grifols	Alphanine	4,924,330
Novo Nordisk	NovoSeven (mg)	5,018
Octapharma	Nanotiv	436,000
Pfizer	BeneFix	70,654,127

# Market Share of factor IX concentrates known to have been used by UK Haemophilia Centres



## Products used in the treatment of von Willebrand's patients

Manufacturer	Product	Total Units
	DDAVP	45
	Desmopressin-DDAVP (4mcg/ml)	2,002
	Desmopressin-Octim nasal spray (150mcg/2.5ml vial)	74,672
	Desmopressin-Octim Sub-cut (15mcg/ml)	9,608
<b>Baxter</b>	Advate	83,974
<b>Bayer</b>	Kogenate	28,500
<b>BPL</b>	BPL FVIII	137,655
<b>CSL Behring</b>	Haemate P	14,627,600
	Haemocompletten P (g)	4
<b>Grifols</b>	Alphanate	2,578,880
	Fanhdi	46,000
	Wilfactin	540,350
<b>Novo Nordisk</b>	NovoSeven (mg)	450
<b>Octapharma</b>	Wilate	279,900
<b>Pfizer</b>	Refacto AF	1,500

## Von Willebrand's Sub-Types

vW Sub-types currently in register	Number of patients
1	2231
1A	4
1B	2
2	103
2A	171
2B	70
2D	1
2M	85
Normandy (2N)	42
3	72
Vincenza	2
Other	30
Type unreported	6066
<b>Total</b>	<b>8879</b>

## Acquired defects: Materials used by number of patients

Product	Acquired Haemophilia A	Acquired von Willebrands	Acquired FX Deficiency	Total
Advate	4	1	-	5
Desmopressin-Octim Sub-cut (15mcg/ml)	1	-	-	1
Feiba	53	1	1	55
Haemate P	-	13	-	13
Haemocompletten P (g)	2	-	-	2
Helixate Nexgen	2	-	-	2
Kogenate	1	-	-	1
NovoSeven (mg)	22	1	-	23
Octaplex	1	-	-	1
Refacto	1	-	-	1

## Adverse events

Adverse Event	Number of Events
New Inhibitor	14
Non-A, Non-B or Hepatitis C Transmission	0
Other Events *	2
Transfusion Reaction	5
<b>Total</b>	<b>21</b>

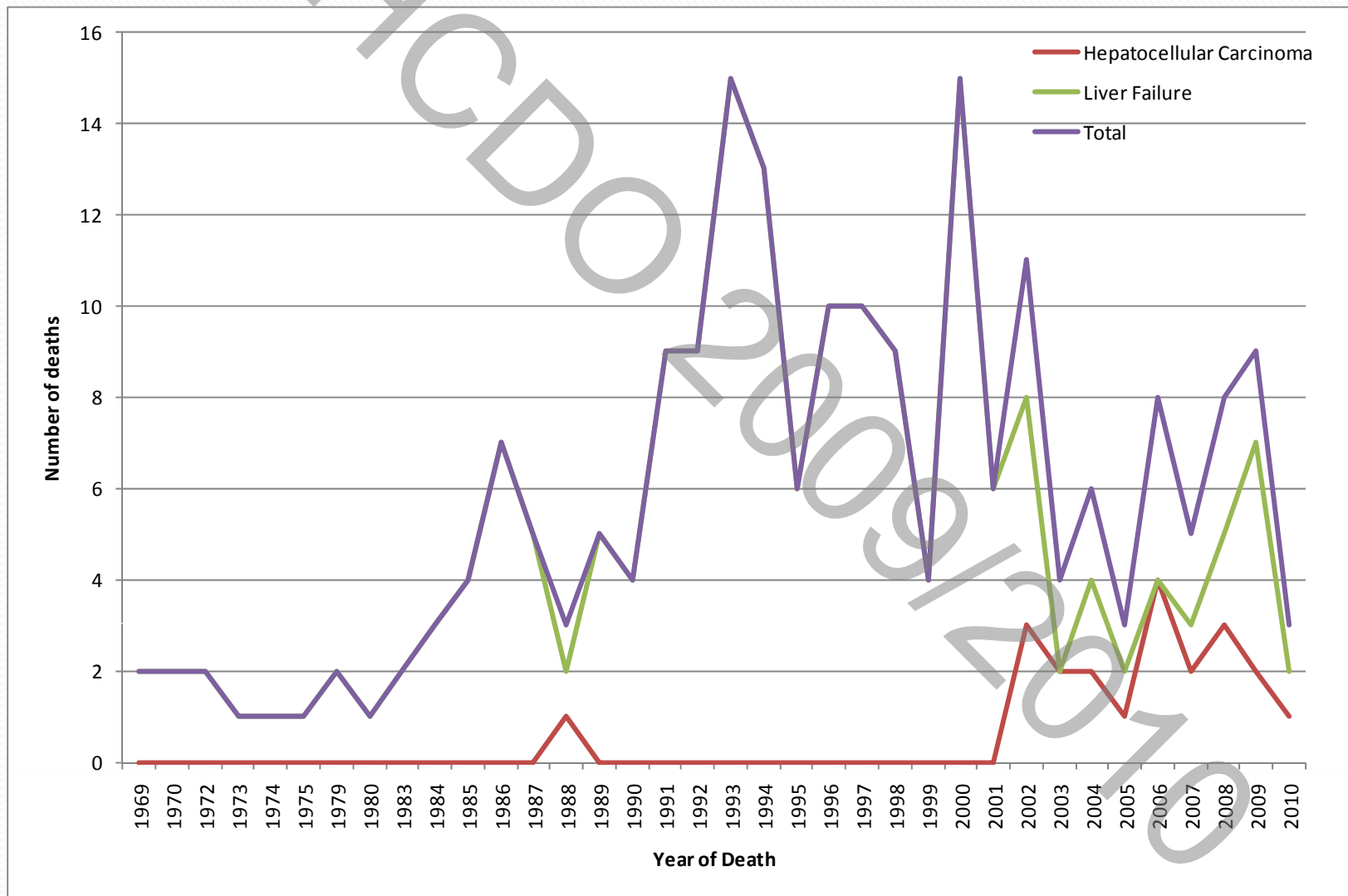
**Estimate of number of patients exposed to hepatitis C, based on historical clotting factor concentrate exposure**

Coagulation Defect	Alive at 31/03/2010	Died by 31/03/2010	Total
Haemophilia A	1,882	1,571	3,453
Haemophilia B	521	192	713
Females with VIII deficiency	79	18	97
Females with IX deficiency	40	5	45
von Willebrand disease	207	68	275
F.VII deficiency	5	-	5
F.X deficiency	7	1	8
F.XI Deficiency	4	3	7
Prothrombin Deficiency	1	-	1
Combined V+VIII Deficiency	2	-	2
Other combined diagnoses	3	2	5
Acquired Haemophilia A	2	24	26
Acquired Haemophilia B	1	1	2
Acquired von Willebrands	1	1	2
Platelet defects	1	-	1
Haemophilia A with Liver Transplant	8	10	18
Haemophilia B with Liver Transplant	2	2	4
Factor X deficiency with Liver Transplant	-	1	1
Miscellaneous	-	1	1
Unclassified	9	-	9
<b>Total</b>	<b>2,775</b>	<b>1,900</b>	<b>4,675</b>

**Annualised UK Deaths  
from Liver Disease 1969-  
2010**

Year of death	Hepatocellular Carcinoma	Liver Failure	Total
1969	-	2	2
1970	-	2	2
1972	-	2	2
1973	-	1	1
1974	-	1	1
1975	-	1	1
1979	-	2	2
1980	-	1	1
1983	-	2	2
1984	-	3	3
1985	-	4	4
1986	-	7	7
1987	-	5	5
1988	1	2	3
1989	-	5	5
1990	-	4	4
1991	-	9	9
1992	-	9	9
1993	-	15	15
1994	-	13	13
1995	-	6	6
1996	-	10	10
1997	-	10	10
1998	-	9	9
1999	-	4	4
2000	-	15	15
2001	-	6	6
2002	3	8	11
2003	2	2	4
2004	2	4	6
2005	1	2	3
2006	4	4	8
2007	2	3	5
2008	3	5	8
2009	2	7	9
2010	1	2	3
<b>Total</b>	<b>21</b>	<b>187</b>	<b>208</b>

## Annualised UK Deaths from Liver Disease 1969-2010



vCJD

Coagulation Defect	Patients at public health risk	Patients known to have received implicated batch(es)
Haemophilia A	2305	565
Haemophilia B	576	171
Females with VIII deficiency	59	2
Females with IX deficiency	55	3
von Willebrand disease	563	39
F.VII deficiency	35	-
F.X deficiency	29	8
F.XI Deficiency	119	-
F.XII (Hageman) defect	2	-
F.XIII Deficiency	22	-
Fibrinogen Deficiency	2	-
Combined II+VII+IX+X Deficiency	1	-
Combined V+VIII Deficiency	7	-
Antithrombin III	11	-
Acquired Haemophilia A	32	5
Acquired Haemophilia B	1	1
Acquired von Willebrands	6	1
Acquired F.XIII Deficiency	1	-
Severe Platelet Disorders - Other	6	-
Haemophilia A with Liver Transplant	10	2
Haemophilia B with Liver Transplant	3	1
Misc.	1	-
Unclassified	14	1
Temporary coagulation defect, now normal	3	-
Combined diagnoses	8	1
<b>Total</b>	<b>3871</b>	<b>800</b>

## Report on the enhanced surveillance of patients 'at risk' of CJD

### Description of age-groups at exposure by exposure group

At risk group	Age group at exposure <sup>1</sup>									Total	No DOB
	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79	>80		
Recipients of implicated products	177	189	158	111	86	41	21	11	5	799	1
Recipients of non-implicated products	1,176	636	462	384	232	139	36	6	0	3,071	1

### Date of exposure by exposure group

At risk group	Total patients by year group of exposure <sup>1</sup>							Missing year group	Total
	1980-1984	1985-1989	1990-1994	1995-1999	2000-2004	2005-2009	2010		
Recipients of implicated products	0	57	336	407	0	0	0	0	800
Recipients of non-implicated products	2,598	249	176	46	1	1	0	1	3,072

## Report on the enhanced surveillance of patients 'at risk' of CJD (Cont'd)

### Description of genotype and sex by exposure groups

Exposure group	Gender		
	Male	Female	Not known
Recipients of implicated products	761	38	1
Recipients of non-implicated products	2,525	547	0

### Description of genotype and sex by exposure groups

Exposure group	Total at risk	Total deaths	Person Years at risk <sup>1</sup>			Total post mortems done
			Total	Median	Range	
Recipients of implicated products	800	73	12,783	15.8	0-23	8
Recipients of non-implicated products	3,072	343	86,271	31.0	1-31	50

## Report on the enhanced surveillance of patients 'at risk' of CJD (Cont'd)

Age-group distribution of patients who are currently alive by exposure group

At risk group	Current Age group of living at risk patients (maximum alive)										Total	No DOB
	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79	80-89	≥ 90		
Recipients of implicated products	0	42	201	166	144	84	63	18	7	1	<b>726</b>	1
Recipients of non-implicated products	2	184	472	438	568	417	346	195	88	18	<b>2,728</b>	1

Number of years lived following exposure for patients currently alive

Number of years since exposure	Current age group of living patients (maximum alive)									Total alive
	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79	80-89	
0-4	0	0	0	0	0	0	0	0	0	<b>0</b>
5-9	2	0	0	0	0	0	0	0	0	<b>2</b>
10-14	0	51	62	53	45	25	18	4	2	<b>260</b>
15-19	0	175	119	90	72	42	37	13	5	<b>553</b>
≥20	0	0	492	461	595	434	354	196	107	<b>2639</b>

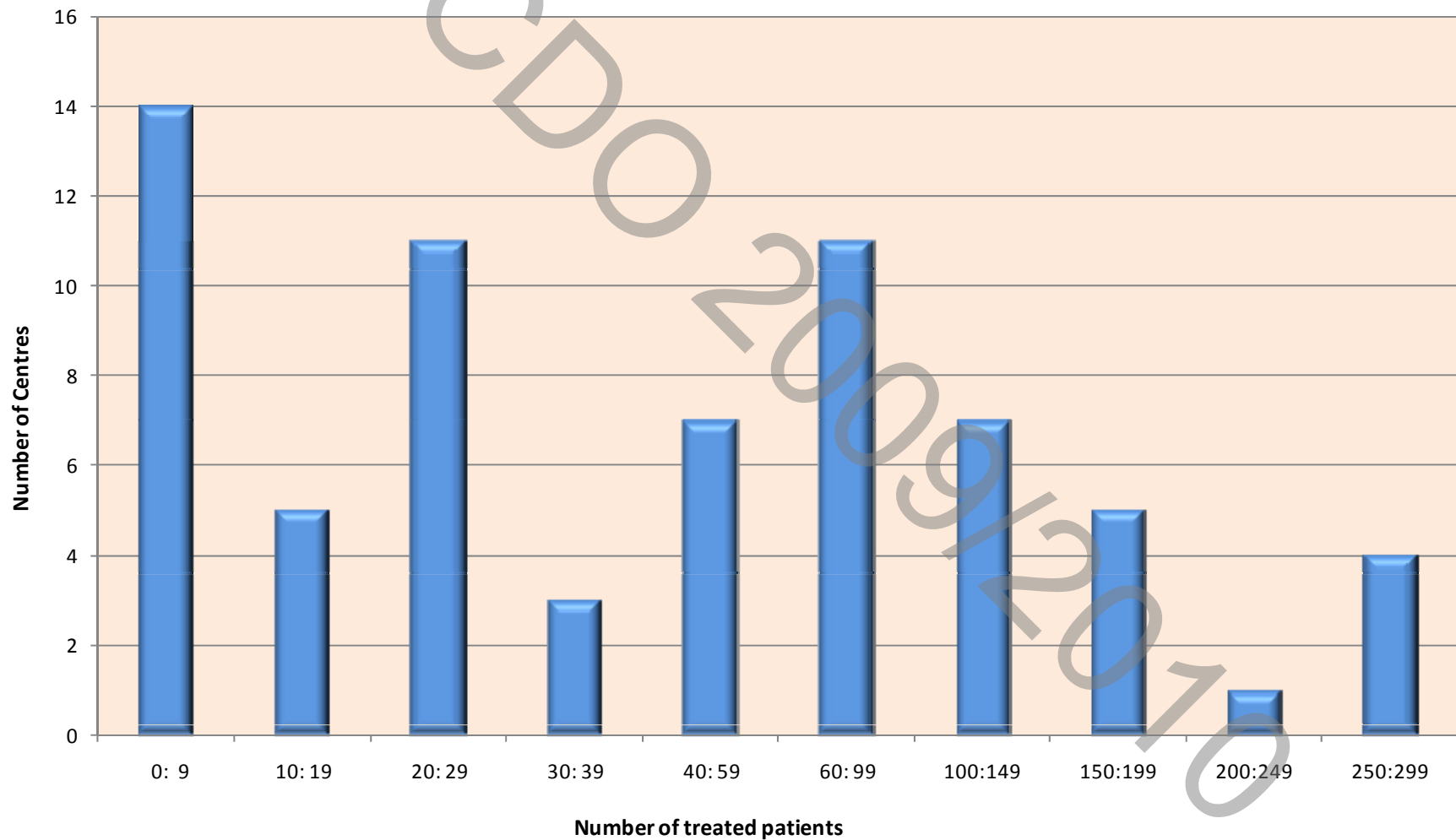
**Causes of death in  
Haemophilia A, B, Females  
with FVIII/FIX deficiency & von  
Willebrand's disease patients**

Cause of Death	Severity		
	Severe	Moderate	Mild
AIDS	-	1	-
Carcinoma	1	1	18
Encephalopathy	1	-	-
Epilepsy	-	-	1
Haemorrhage	4	1	7
Hepatocellular Carcinoma	1	2	-
Infection (Bacterial)	2	3	10
Ischaemic Heart Disease	2	1	5
Liver Failure	2	-	1
Lymphoproliferative Malignancy	-	-	1
Renal Failure	2	-	1
Stroke (thrombotic)	-	-	1
Stroke (Unknown)	-	-	2
Suicide	-	1	-
Unknown	1	-	10
<b>Total</b>	<b>16</b>	<b>10</b>	<b>57</b>

## Causes of death - Rarer Defects

Coagulation Defect	Cause of Death	Number of deaths
Acquired Haemophilia A	Carcinoma	1
	COAD	1
	Haemorrhage (Misc)	1
	Ischaemic Heart Disease	3
	Renal Failure	1
	Stroke (Unknown)	1
	Unknown	9
Acquired von Willebrands	Carcinoma	1
Dysfibrinogenaemia	Unknown	1
F.VII deficiency	COAD	1
F.X deficiency	Ischaemic Heart Disease	1
F.XI Deficiency	Carcinoma	3
	Infection (Bacterial)	2
	Unknown	1
Fibrinogen Deficiency	Infection (Bacterial)	1
	Unknown	1
Glanzmanns Thrombasthenia	Accident	1
Haemophilia A with Liver Transplant	Liver Failure	1
	Lymphoproliferative Malignancy	1
Combined diagnoses	Cerebral haemorrhage	1
	Ischaemic Heart Disease	2
Platelet defects (misc)	Infection (Bacterial)	1
	Ischaemic Heart Disease	1
	Liver Failure	1
	Peripheral vascular disease	1
<b>Total</b>		<b>39</b>

## Total number of patients with Haemophilia A, Haemophilia B or von Willebrand's Disease treated by UK Haemophilia Centres



## Total number of severely affected patients with Haemophilia A, Haemophilia B or von Willebrand's Disease treated by UK Haemophilia Centres

