

# UKHCDO ANNUAL REPORT

## Bleeding Disorder Statistics for the Financial Year 2008/2009

Data Source: National Haemophilia Database,  
September 2009

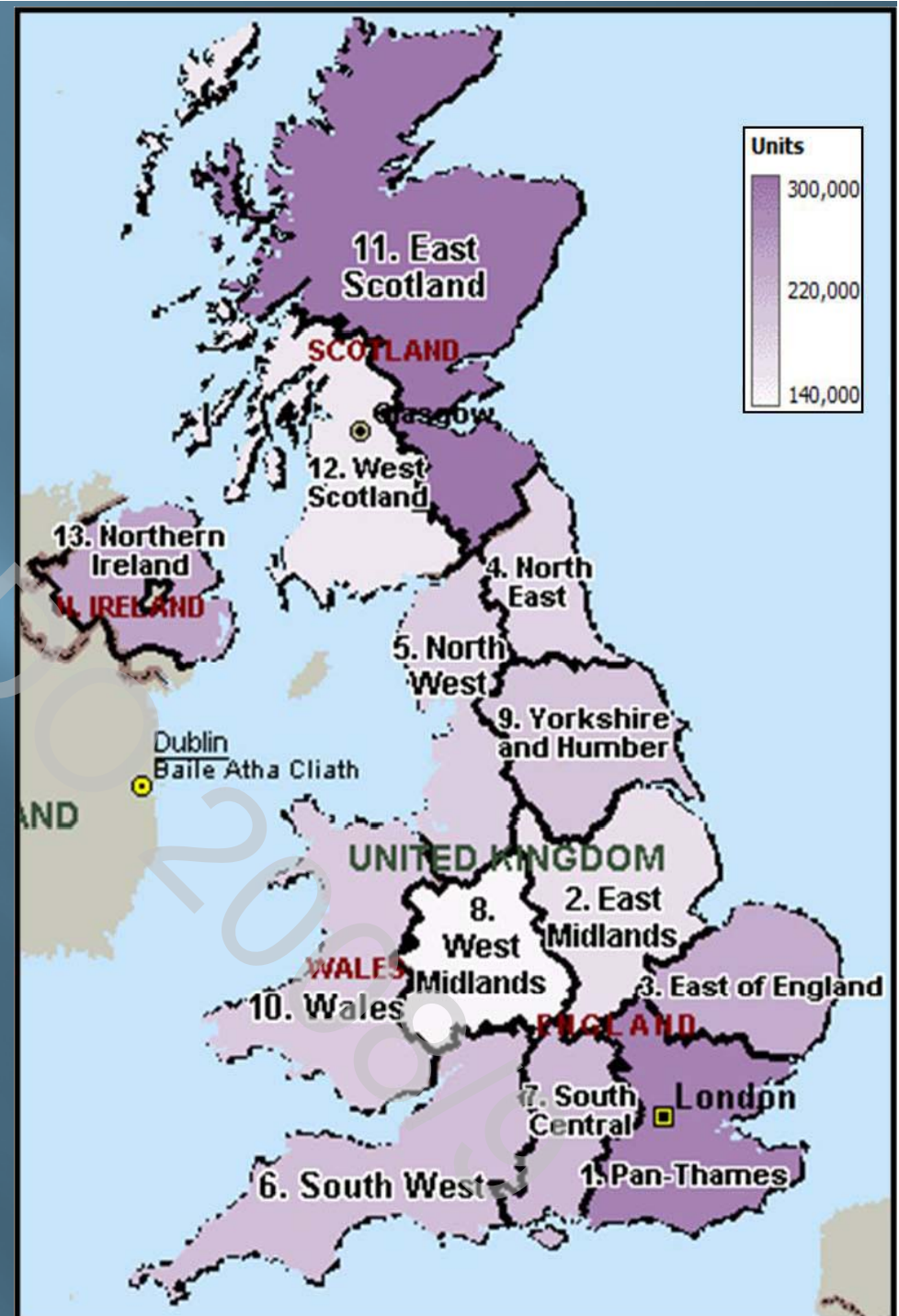
## New Registrations of Haemophilia A & B between April 2008 & March 2009, by age at the end of March 2009 and disease severity

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	13	35	6	89
	10 : 19	4	1	9	2	16
	20 : 29	6	1	5	0	12
	30 : 39	0	2	5	0	7
	40 : 49	1	0	3	1	5
	50 - 59	0	0	12	0	12
	60 : 69	0	0	7	0	7
	70 : 79	0	0	5	1	6
	80 : 89	0	0	1	0	1
<b>Total</b>		<b>46</b>	<b>17</b>	<b>82</b>	<b>10</b>	<b>155</b>
Haemophilia B	0 : 9	6	3	5	2	16
	10 : 19	0	2	1	0	3
	20 : 29	2	1	0	0	3
	30 : 39	0	1	2	0	3
	70 : 79	0	0	1	0	1
<b>Total</b>		<b>8</b>	<b>7</b>	<b>9</b>	<b>2</b>	<b>26</b>

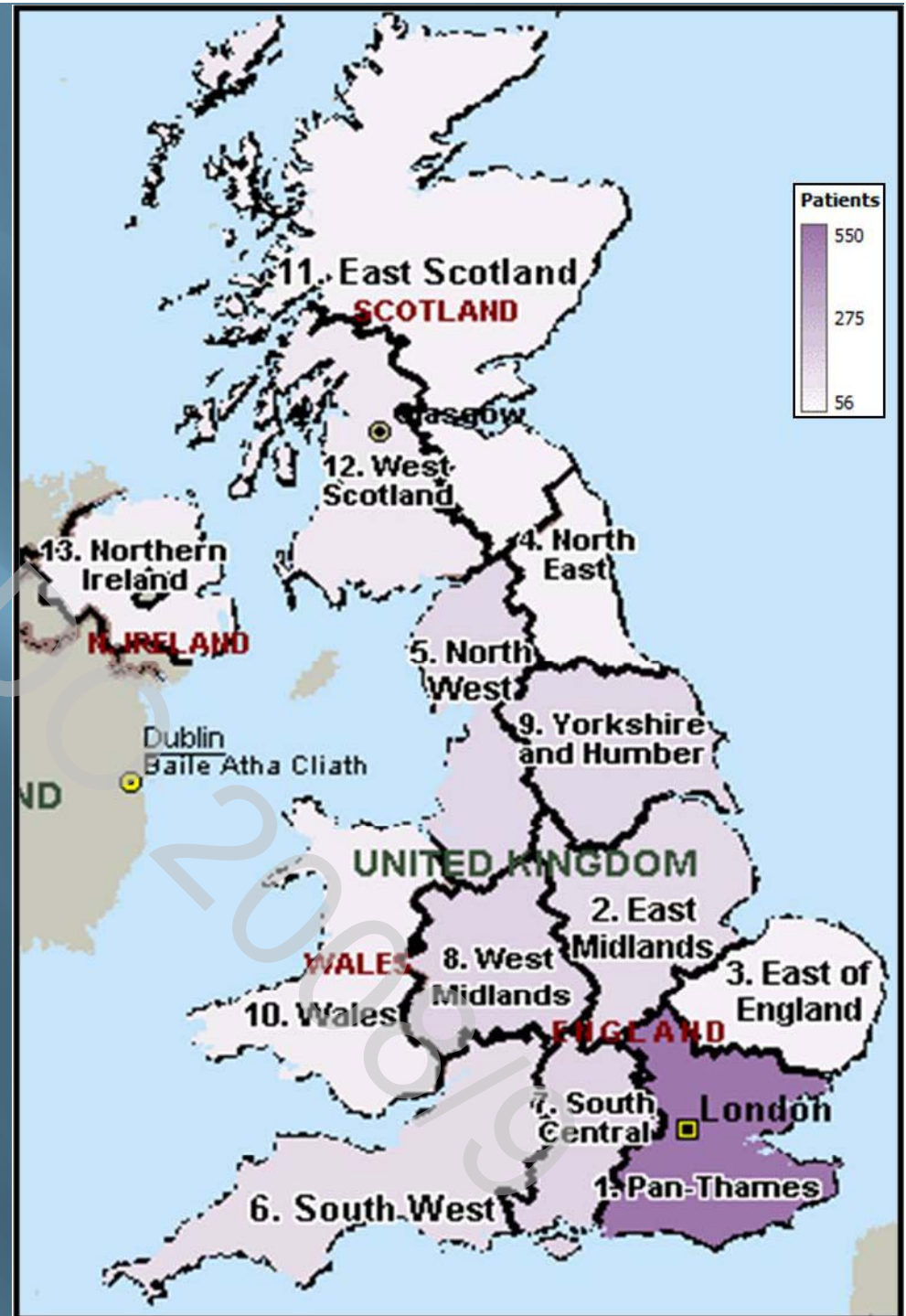
In Register – The total number of haemophilia A, B and von Willebrand's disease patients currently in the register, by severity, age group and gender

Diagnosis	Males <18					Males =>18					Females <18					Females =>18					Overall total
	≤ 1	>1 & <5	≥ 5	NK	Sub Total	≤ 1	>1 & <5	≥ 5	NK	Sub Total	≤ 1	>1 & <5	≥ 5	NK	Sub Total	≤ 1	>1 & <5	≥ 5	NK	Sub Total	
Haemophilia A	622	177	663	13	<b>1475</b>	1514	496	2746	243	<b>4999</b>	0	1	0	0	<b>1</b>	2	1	2	0	<b>5</b>	<b>6480</b>
Haemophilia B	114	55	99	5	<b>273</b>	331	208	498	59	<b>1096</b>	0	0	1	0	<b>1</b>	0	1	1	0	<b>2</b>	<b>1372</b>
von Willebrand disease	17	24	775	91	<b>907</b>	49	48	2386	104	<b>2587</b>	7	27	731	96	<b>861</b>	51	67	4481	300	<b>4899</b>	<b>9254</b>

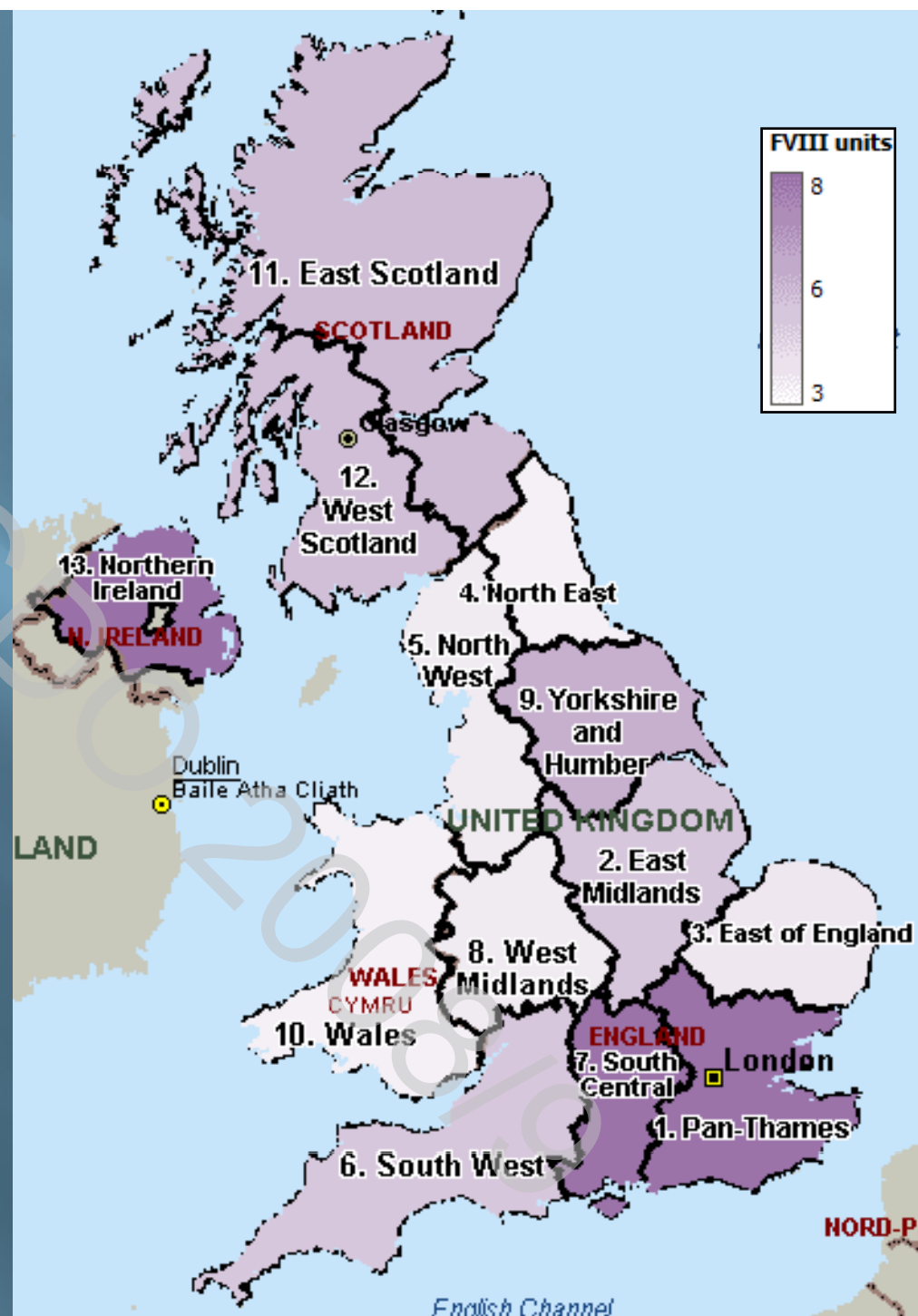
Factor VIII Usage (inc. inhibitor)  
divided by number of Severe  
Haemophilia A patients per  
territory



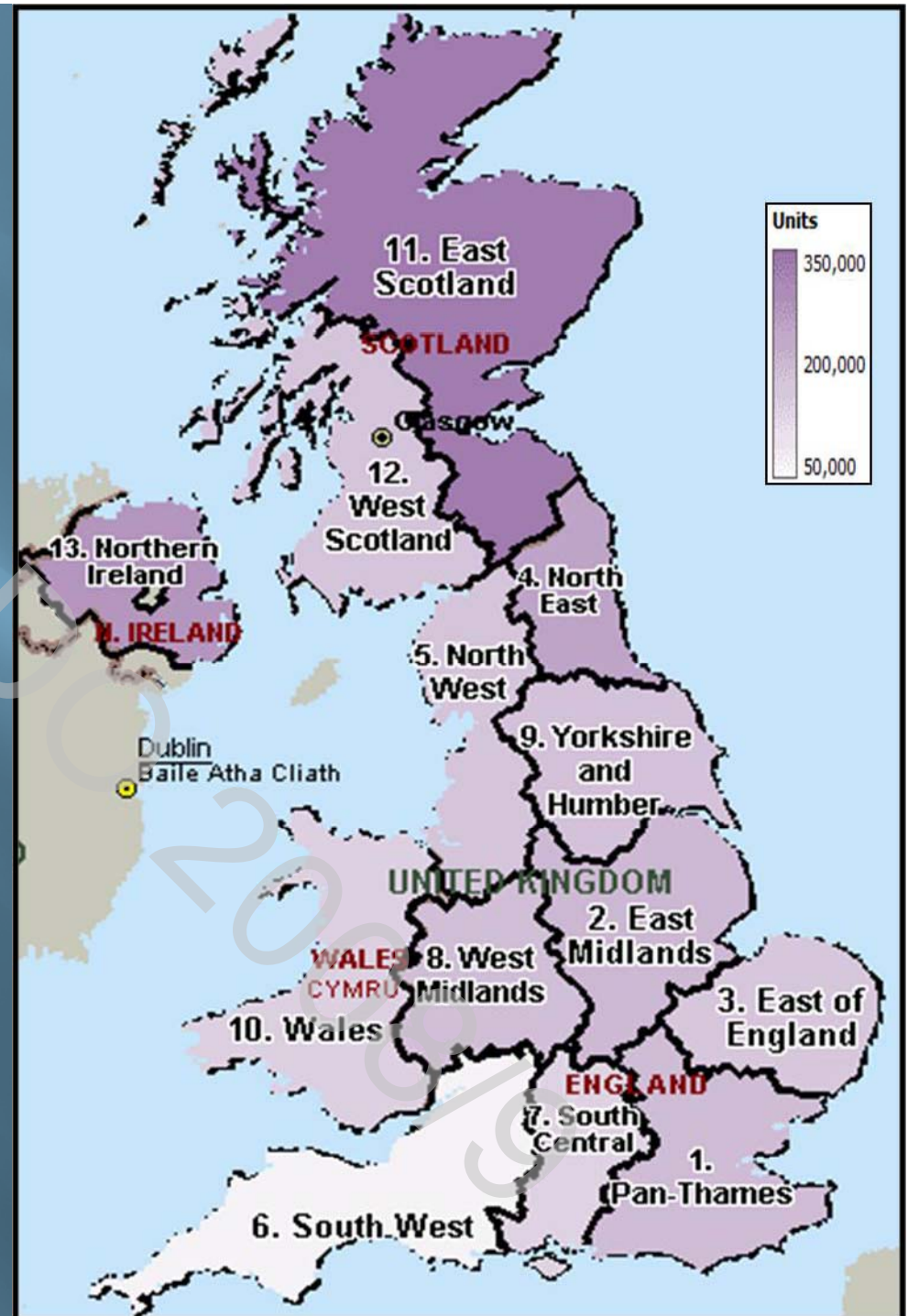
# Total Number of Severe Haemophilia A & B Patients by Territory



# FVIII Units - usage per capita of population



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

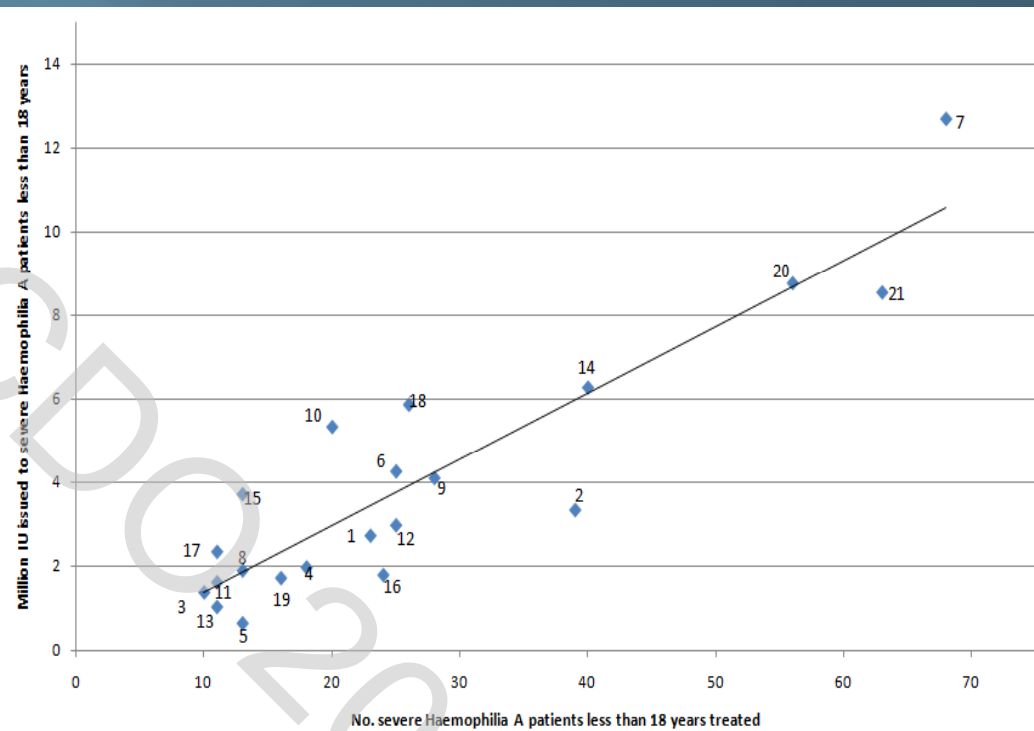


## Factor VIII & FIX Usage by territory for Severe Haemophilia A & B patients (inc. inhibitor)

Specialised Commissioning Group / Region	Severe HpA Patients	Severe HpB patients	Total Severe HpA & HpB Patients	Total FVIII Units Used	Total FIX Units Used	Mean Usage Haemophilia A	Mean Usage Haemophilia B	FVIII Units Per Capita
1. Pan-Thames Haemophilia Consortium	416	112	528	117,171,461	21,604,790	281,662	192,900	7.87
2. East Midlands SCG	119	24	143	21,079,679	4,727,105	177,140	196,963	4.93
3. East of England SCG	37	19	56	8,519,373	3,135,118	230,253	165,006	3.84
4. North East SCG	47	11	58	8,879,159	2,653,000	188,918	241,182	3.49
5. North West SCG	130	22	152	25,550,910	3,791,325	196,545	172,333	3.74
6. South West SCG	118	18	136	25,126,333	1,301,066	212,935	72,281	4.99
7. South Central SCG	142	21	163	31,143,256	2,973,190	219,319	141,580	7.94
8. West Midlands SCG	134	29	163	19,939,719	5,302,346	148,804	182,840	3.74
9. Yorkshire and Humber SCG	142	17	159	28,944,015	3,066,587	203,831	180,387	5.74
10. Wales	52	12	64	10,295,887	1,740,000	197,998	145,000	3.50
11. East Scotland	51	4	55	15,037,983	1,303,860	294,862	325,965	5.37
12. West Scotland	66	20	86	11,049,970	3,407,126	167,424	170,356	5.26
13. Northern Ireland	56	6	62	13,473,243	1,635,482	240,594	272,580	7.99

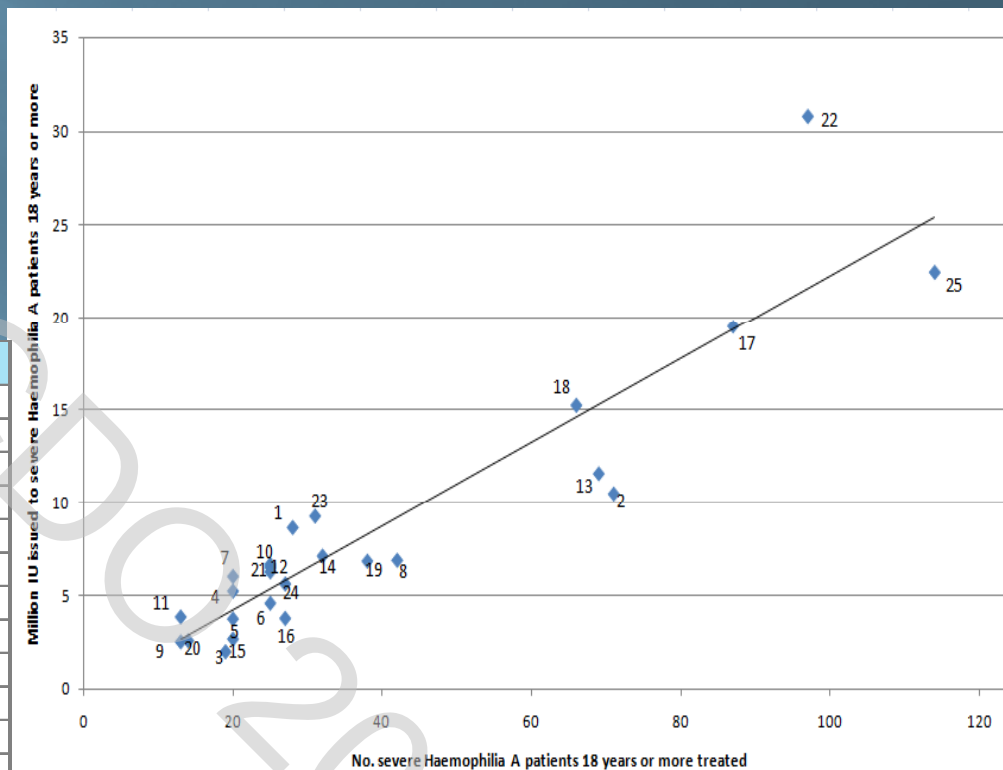
# Severe Haemophilia A - Number of patients with no current inhibitor aged less than 18 years, treated at each centre compared to number of factor VIII units issued by each centre

Key	Centre	Patients	Units	Average
1	Belfast Children's	23	2,726,765	118,555
2	Birmingham (Children's)	39	3,338,000	85,590
3	Bristol (Infirmary & Children's)	10	1,379,000	137,900
4	Cambridge	18	1,971,623	109,535
5	Cardiff	13	642,750	49,442
6	Glasgow (RHSC)	25	4,260,474	170,419
7	Great Ormond Street, London	68	12,691,363	186,638
8	Hammersmith Hospital, London	13	1,898,000	146,000
9	Leeds	28	4,101,000	146,464
10	Liverpool (Children's)	20	5,337,500	266,875
11	Manchester (Children's)	11	1,619,000	147,182
12	Newcastle uponTyne	25	2,974,500	118,980
13	Nottingham	11	1,031,645	93,786
14	Oxford	40	6,275,500	156,888
15	Royal Free, London	13	3,716,067	285,851
16	Sheffield (Children's)	24	1,790,750	74,615
17	St George's Hospital, London	11	2,343,750	213,068
18	St Thomas' and Guy's Hospitals, London	26	5,867,250	225,663
19	The Royal London	16	1,719,250	107,453
20	Centres with <=4 Patients Meeting Criteria	56	8,762,704	156,477
21	Centres with 5-9 Patients Meeting Criteria	63	8,543,120	135,605

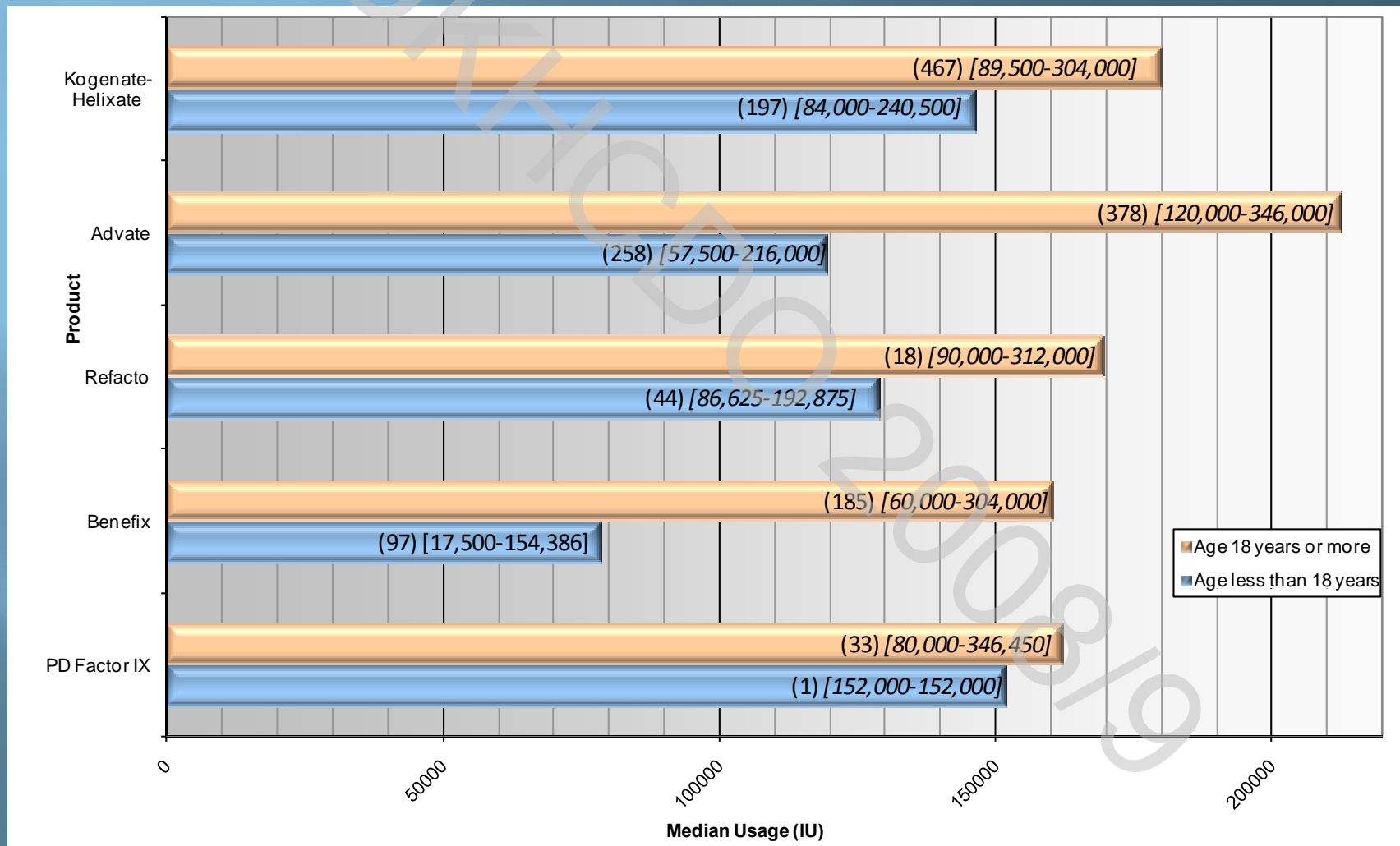


# Severe Haemophilia A - Number of patients with no current inhibitor aged 18 years or more, treated at each centre compared to number of factor VIII units issued by each centre

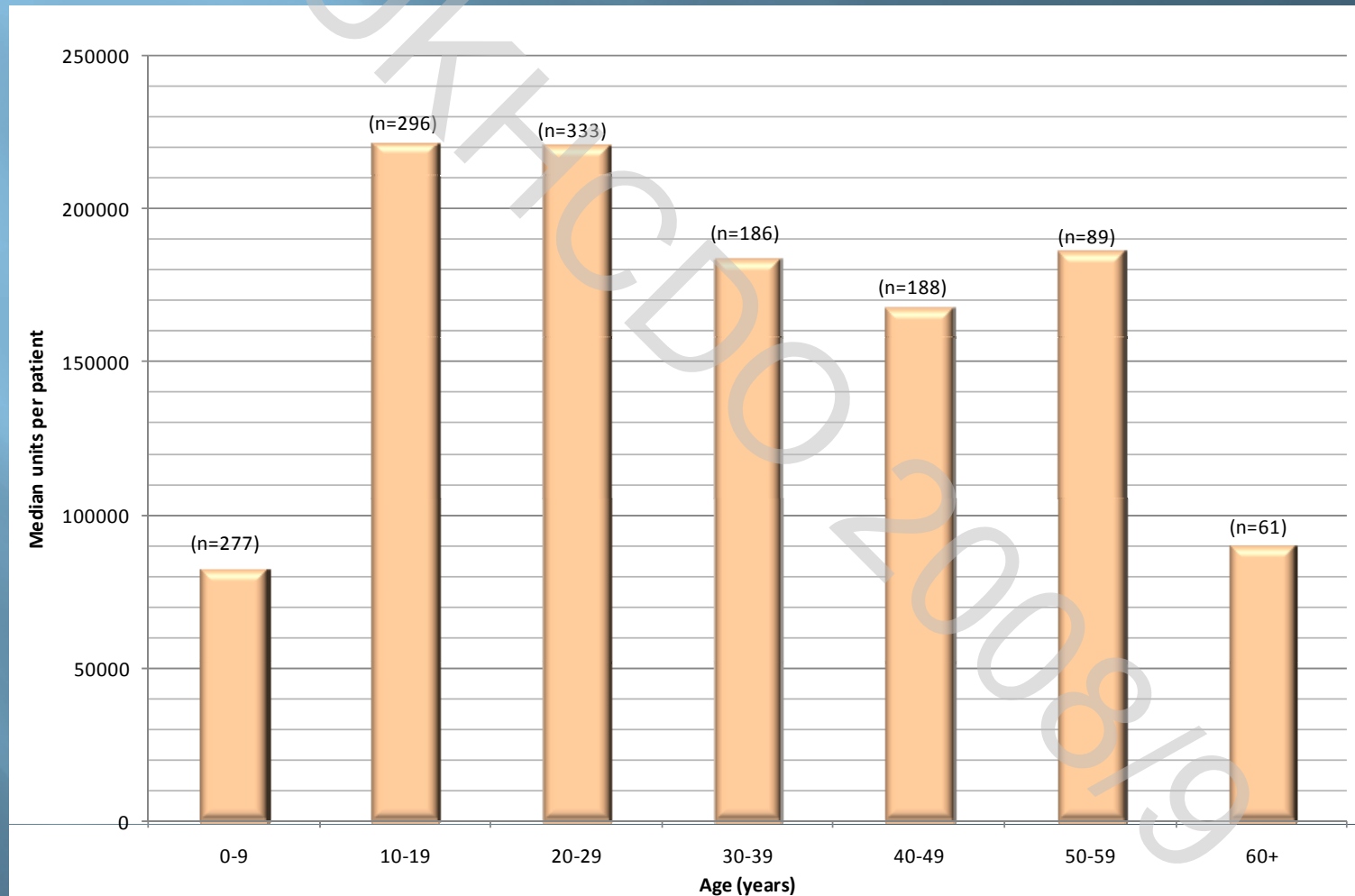
Key	Centre	Patients	Units	Average
1	Belfast Adult's	28	8,679,634	309,987
2	Birmingham (Queen Elizabeth's)	71	10,508,000	148,000
3	Bristol (Infirmary & Children's)	19	2,050,000	107,895
4	Cambridge	20	5,281,661	264,083
5	Canterbury	20	3,800,500	190,025
6	Cardiff	25	4,635,500	185,420
7	Edinburgh	20	6,068,887	303,444
8	Glasgow (RI)	42	6,928,772	164,971
9	Hammersmith Hospital, London	13	2,576,000	198,154
10	Leeds	25	6,713,000	268,520
11	Leicester	13	3,905,000	300,385
12	Liverpool (RI)	25	6,554,160	262,166
13	Manchester (RI)	69	11,593,502	168,022
14	Newcastle uponTyne	32	7,159,000	223,719
15	North Hampshire	20	2,730,000	136,500
16	Nottingham	27	3,829,326	141,827
17	Oxford	87	19,495,082	224,081
18	Royal Free, London	66	15,248,220	231,034
19	Sheffield (Royal Hallamshire)	38	6,883,396	181,142
20	Southampton	14	2,631,500	187,964
21	St George's Hospital, London	25	6,313,061	252,522
22	St Thomas' and Guy's Hospitals, London	97	30,824,500	317,778
23	The Royal London	31	9,293,500	299,790
24	Centres with <=4 Patients Meeting Criteria	27	5,663,832	209,772
25	Centres with 5-9 Patients Meeting Criteria	114	22,472,406	197,126



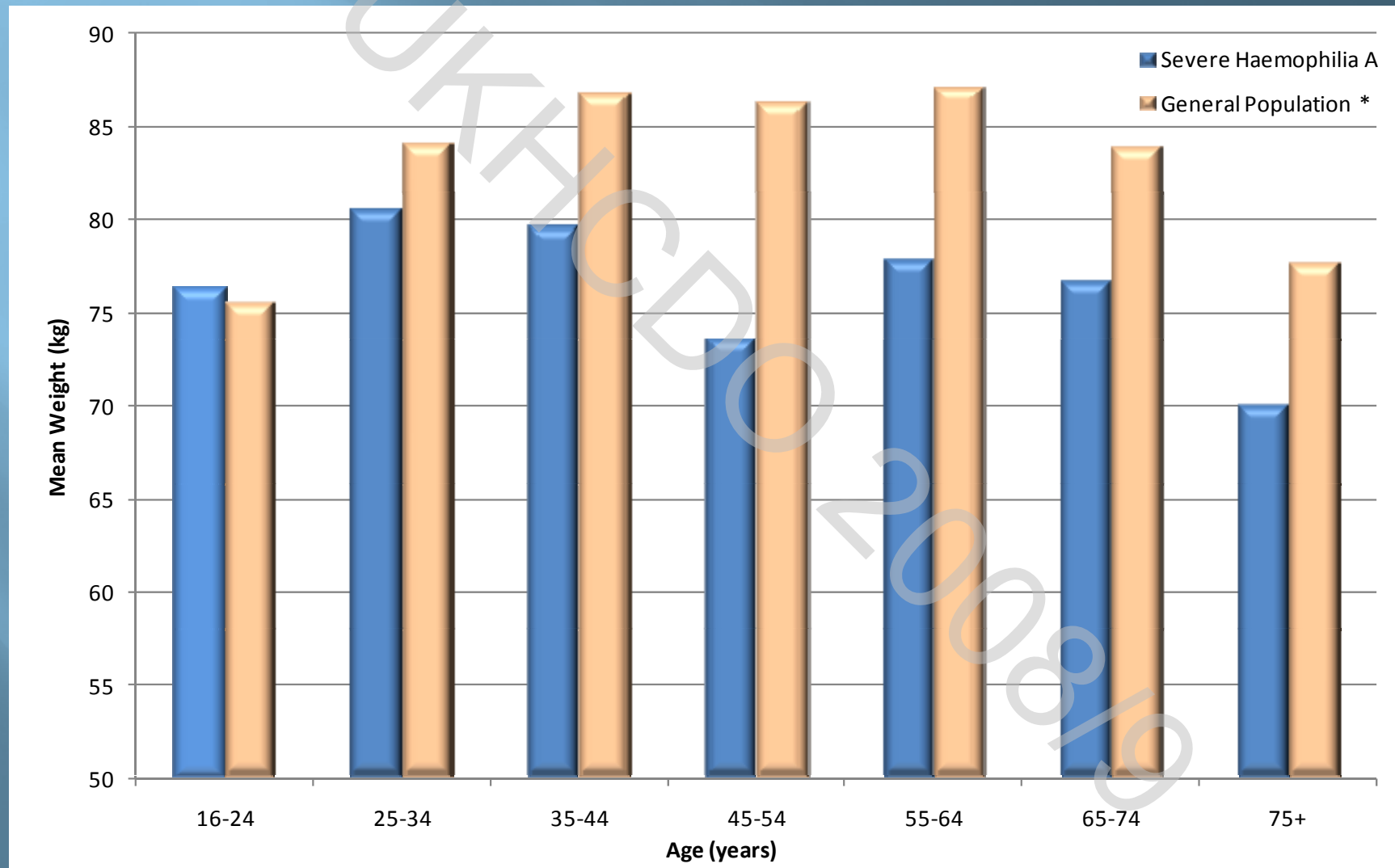
# Severe haemophilia A & B patients with no current inhibitor using only one product between April 2008 & March 2009: median usage



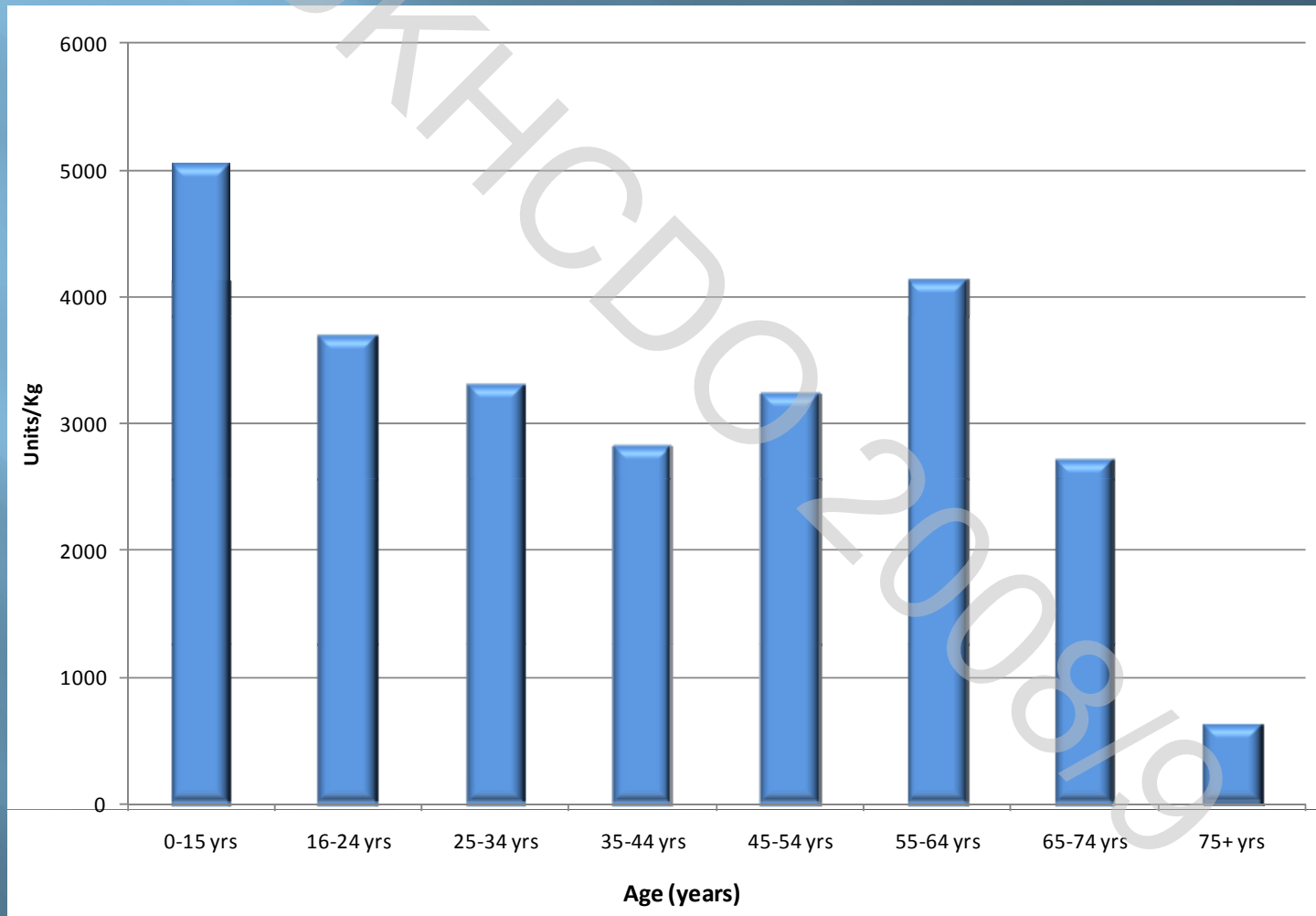
# Severe Haemophilia A patients treated with FVIII products with no current inhibitor: median IU issued



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

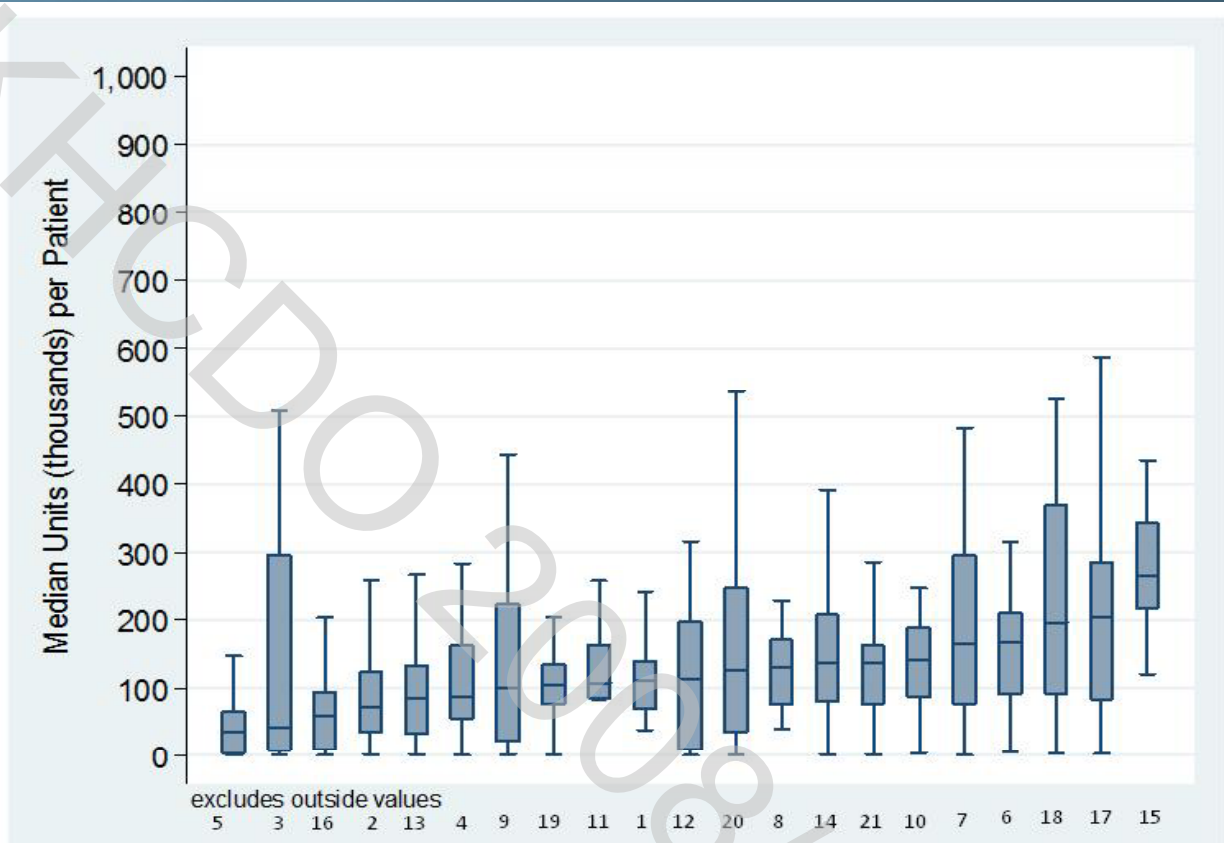


Mean factor VIII units used per kilogram body weight per year in Severe Haemophilia A patients, 2008/09



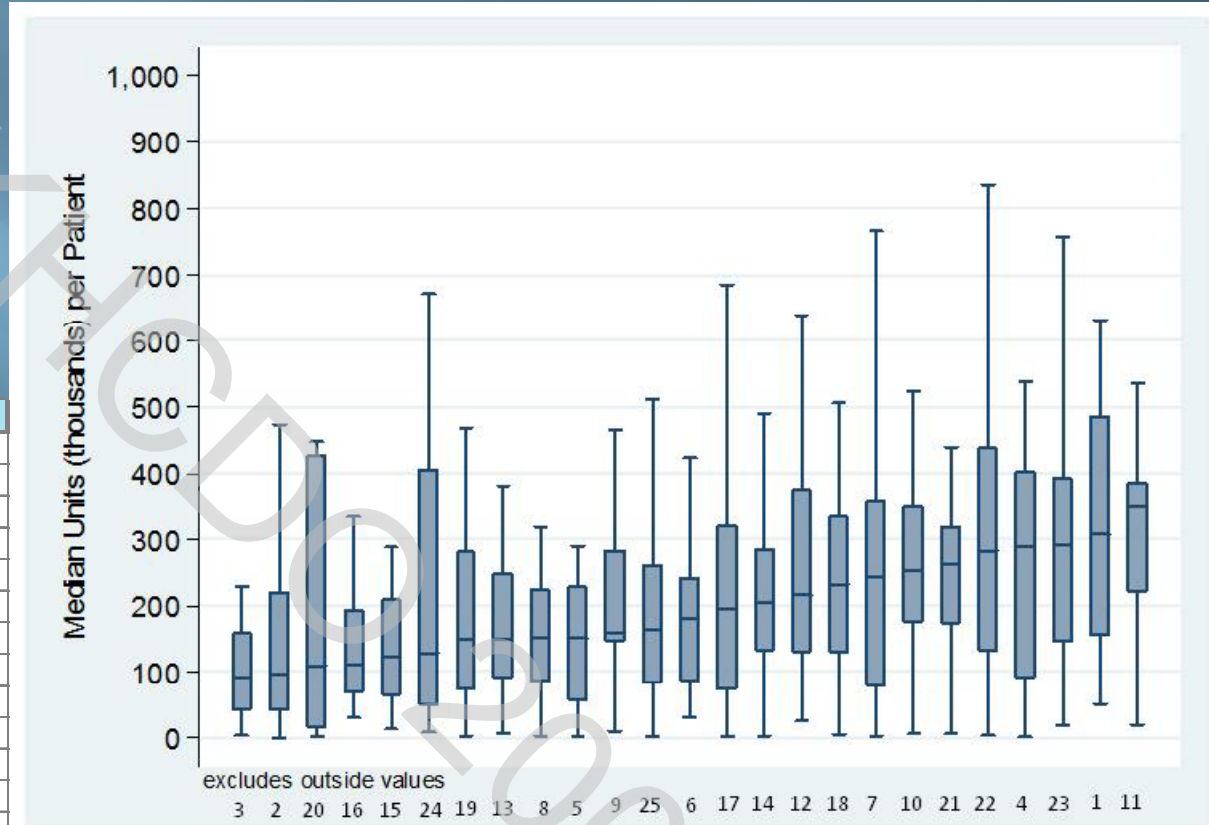
# FVIII usage in severe haemophilia A patients with no current inhibitor by centre, aged less than 18 years

Key	Centre
1	Belfast Children's
2	Birmingham (Children's)
3	Bristol (Infirmary & Children's)
4	Cambridge
5	Cardiff
6	Glasgow (RHSC)
7	Great Ormond Street, London
8	Hammersmith Hospital, London
9	Leeds
10	Liverpool (Children's)
11	Manchester (Children's)
12	Newcastle uponTyne
13	Nottingham
14	Oxford
15	Royal Free, London
16	Sheffield (Children's)
17	St George's Hospital, London
18	St Thomas' and Guy's Hospitals, London
19	The Royal London
20	Centres with <=4 Patients Meeting Criteria
21	Centres with 5-9 Patients Meeting Criteria

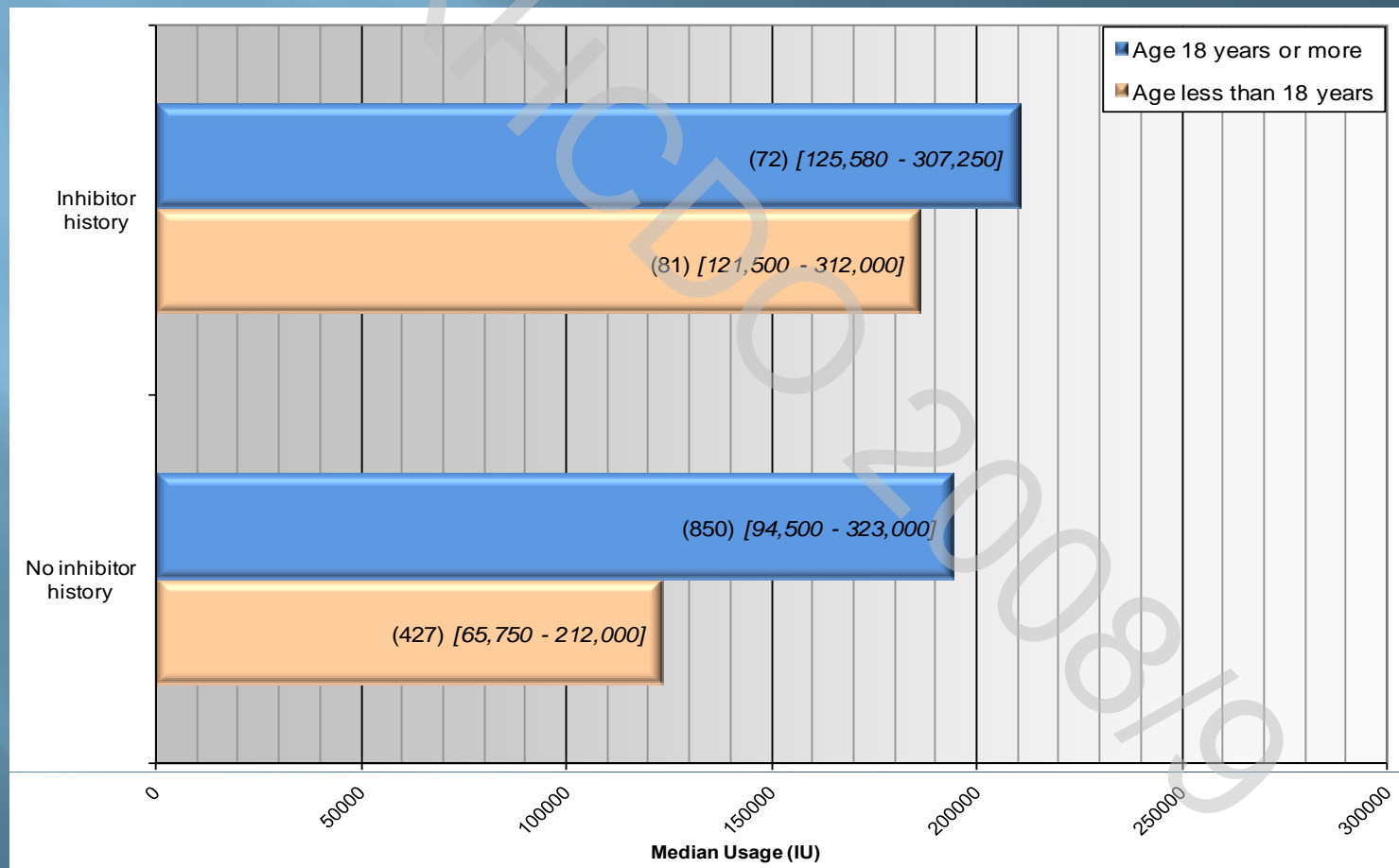


# FVIII usage in severe haemophilia A patients with no current inhibitor by centre, aged 18 years or more

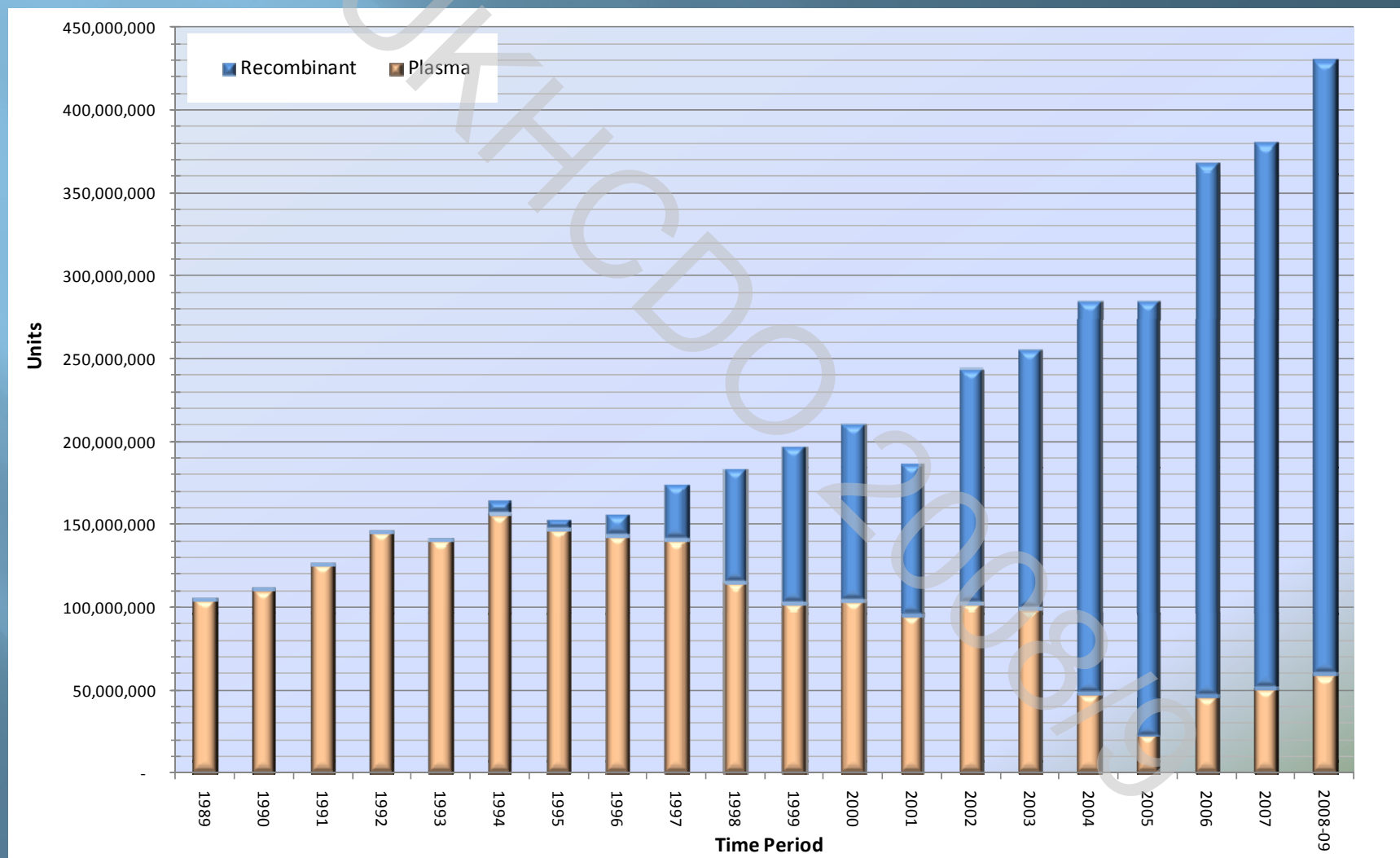
Key	Centre
1	Belfast Adult's
2	Birmingham (Queen Elizabeth's)
3	Bristol (Infirmary & Children's)
4	Cambridge
5	Canterbury
6	Cardiff
7	Edinburgh
8	Glasgow (RI)
9	Hammersmith Hospital, London
10	Leeds
11	Leicester
12	Liverpool (RI)
13	Manchester (RI)
14	Newcastle uponTyne
15	North Hampshire
16	Nottingham
17	Oxford
18	Royal Free, London
19	Sheffield (Royal Hallamshire)
20	Southampton
21	St George's Hospital, London
22	St Thomas' and Guy's Hospitals, London
23	The Royal London
24	Centres with <=4 Patients Meeting Criteria
25	Centres with 5-9 Patients Meeting Criteria



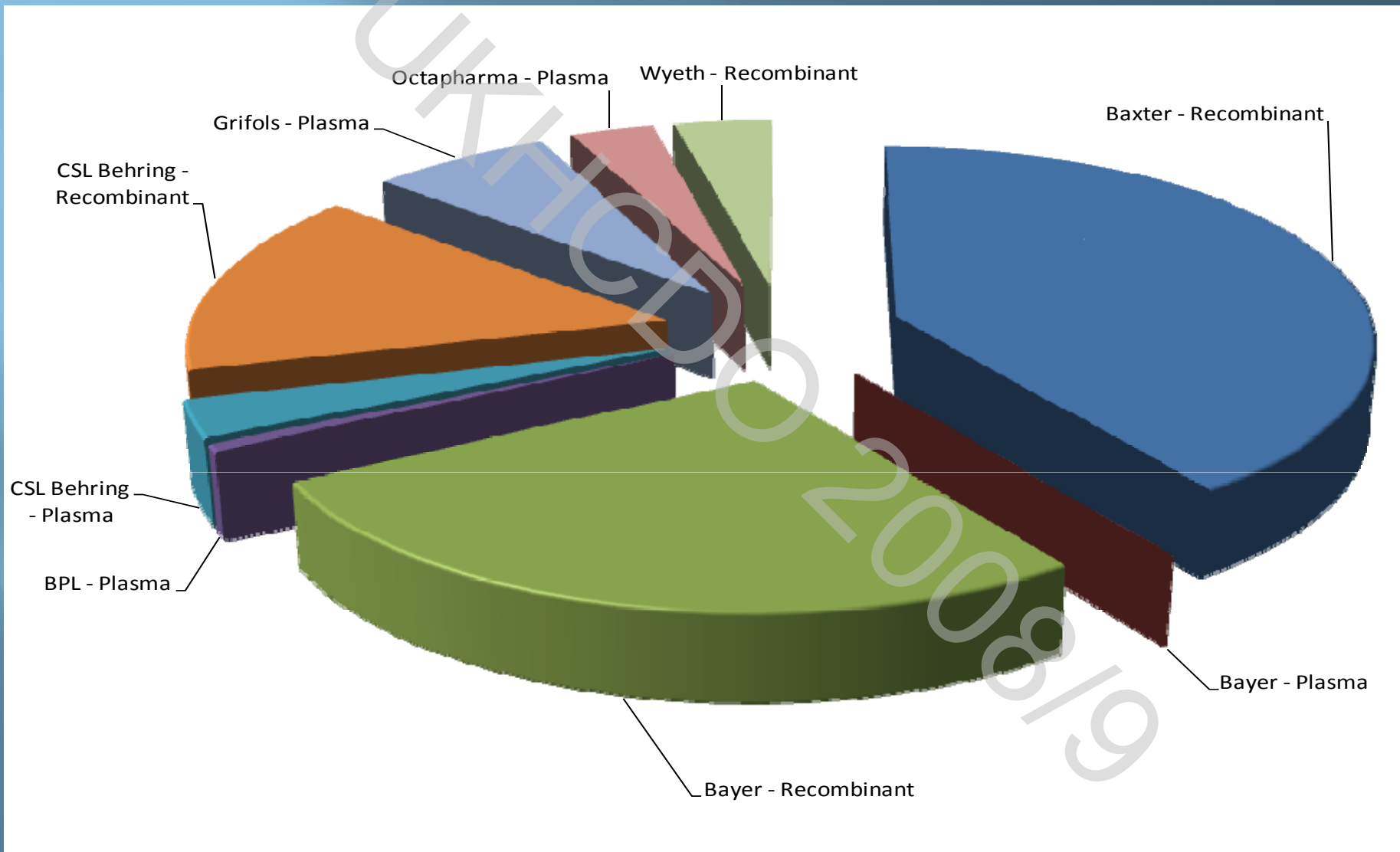
# Severe haemophilia A patients with no current inhibitor between April 2008 & March 2009: median usage by inhibitor history



# Factor VIII usage by UK Haemophilia Centres: between 1989 and 2007 and for financial year 2008-2009



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



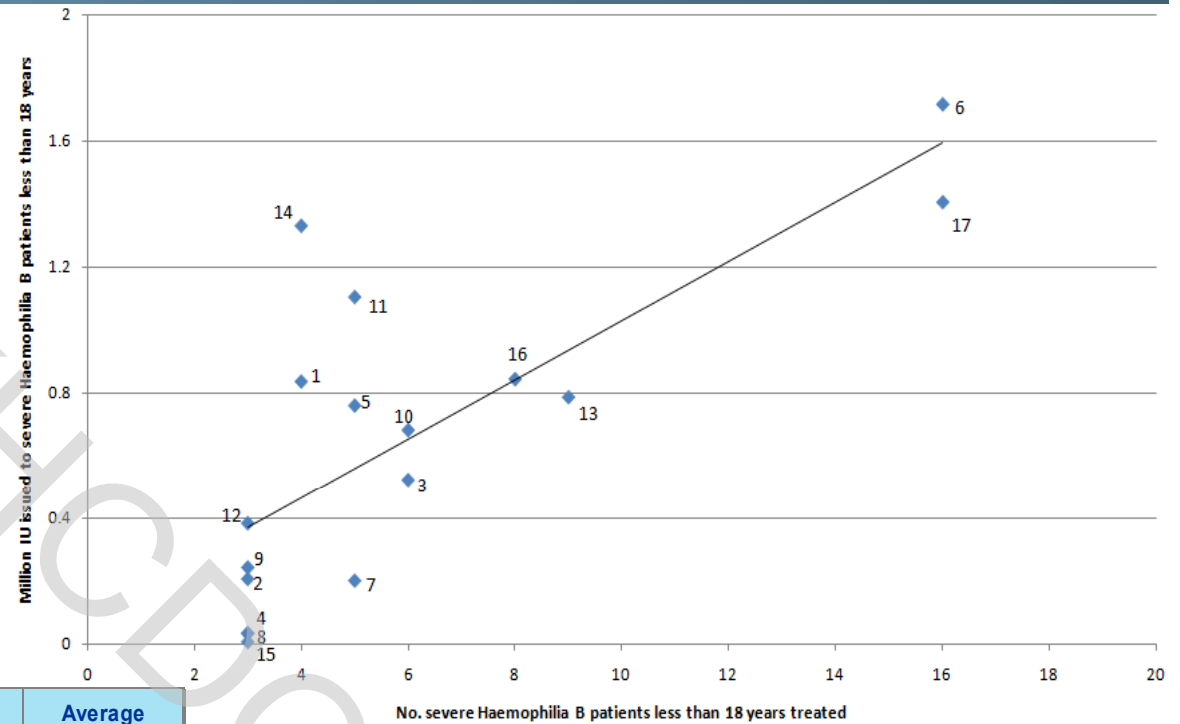
## Prevalence of inhibitors by disease severity

Diagnosis	Number of Patients ever known to have an inhibitor by disease severity														
	≤ 1 iu/dl			>1 and <5 iu/dl			≥ 5 iu/dl			Not Known			Total		
	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%	In Reg	Inhib. Pts	%
Haemophilia A	2138	367	17.17%	675	42	6.22%	3411	54	1.58%	256	1	0.39%	6480	464	7.16%
Haemophilia B	445	11	2.47%	264	0	0.00%	599	2	0.33%	64	1	1.56%	1372	14	1.02%
von Willebrand disease	126	3	2.38%	166	1	0.60%	8376	2	0.02%	597	1	0.17%	9265	7	0.08%

Diagnosis	Patients with a current inhibitor treated between April 2008 and March 2009 by disease severity														
	≤ 1 iu/dl			>1 and <5 iu/dl			≥ 5 iu/dl			Not Known			Total		
	All Treated	Inhib. Pts treated	%	All Treated	Inhib. Pts treated	%	All Treated	Inhib. Pts treated	%	All Treated	Inhib. Pts treated	%	All Treated	Inhib. Pts treated	%
Haemophilia A	1590	156	9.81%	384	16	4.17%	793	25	3.15%	16	2	12.50%	2783	199	7.15%
Haemophilia B	325	8	2.46%	119	0	0.00%	147	2	1.36%	5	1	20.00%	596	11	1.85%
von Willebrand disease	35	1	2.86%	50	1	2.00%	780	4	0.51%	116	0	0.00%	981	6	0.61%

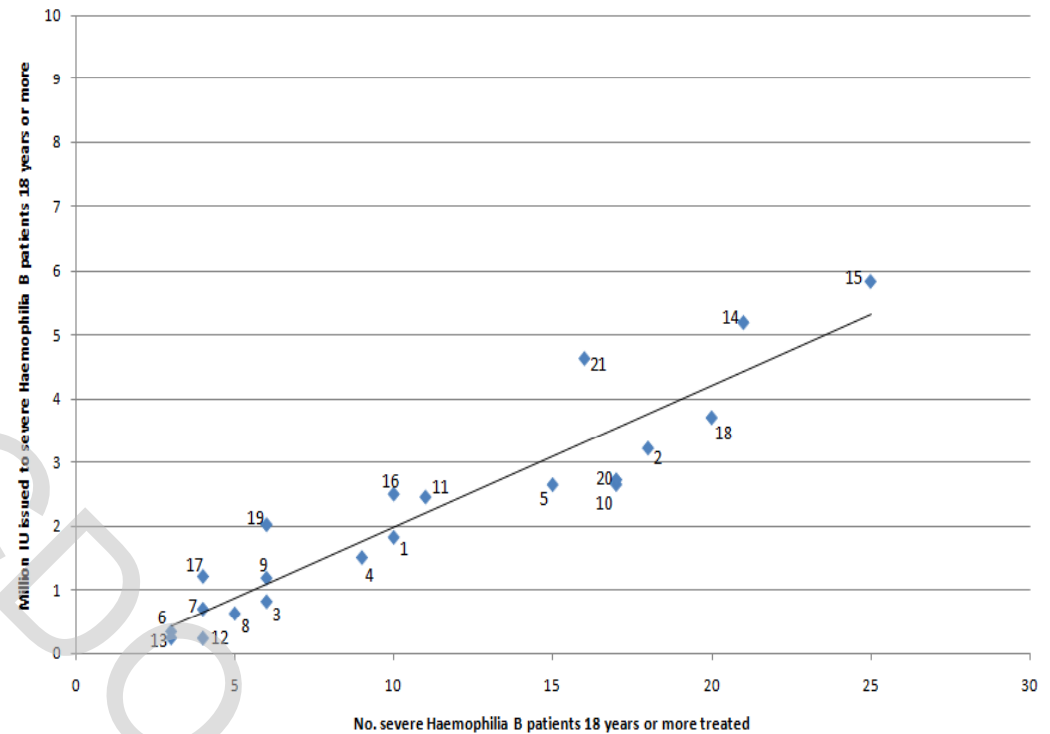
Severe Haemophilia B -  
 Number of patients with no  
 current inhibitor aged less  
 than 18 years, treated at  
 each centre compared to  
 number of factor VIII units  
 issued by each centre



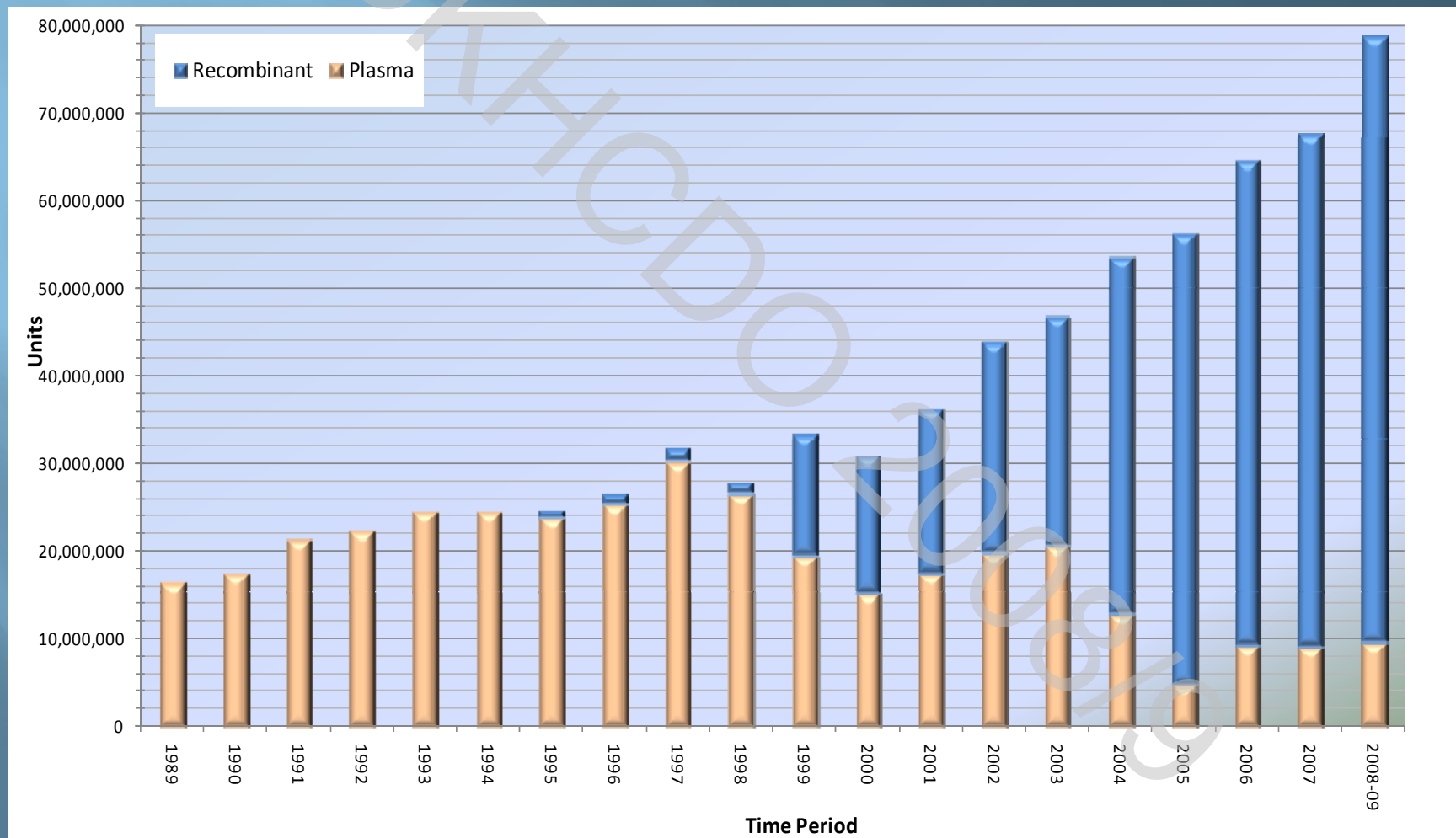
Key	Centre	Patients	Units	Average
1	Belfast Children's	4	834,887	208,722
2	Cambridge	3	204,359	68,120
3	Canterbury	6	521,500	86,917
4	Exeter	3	32,000	10,667
5	Glasgow (RHSC)	5	758,470	151,694
6	Great Ormond Street, London	16	1,714,250	107,141
7	Hammersmith Hospital, London	5	199,010	39,802
8	Kingston upon Hull	3	31,066	10,355
9	Liverpool (Children's)	3	241,000	80,333
10	Manchester (Children's)	6	680,500	113,417
11	Newcastle uponTyne	5	1,103,500	220,700
12	Nottingham	3	381,470	127,157
13	Oxford	9	785,000	87,222
14	Royal Free, London	4	1,329,737	332,434
15	Sheffield (Children's)	3	4,250	1,417
16	Centres with 1 Patient Meeting Criteria	8	842,414	105,302
17	Centres with 2 Patients Meeting Criteria	16	1,404,826	87,802

## Severe Haemophilia B - Number of patients with no current inhibitor aged 18 years or more, treated at each centre compared to number of factor VIII units issued by each centre

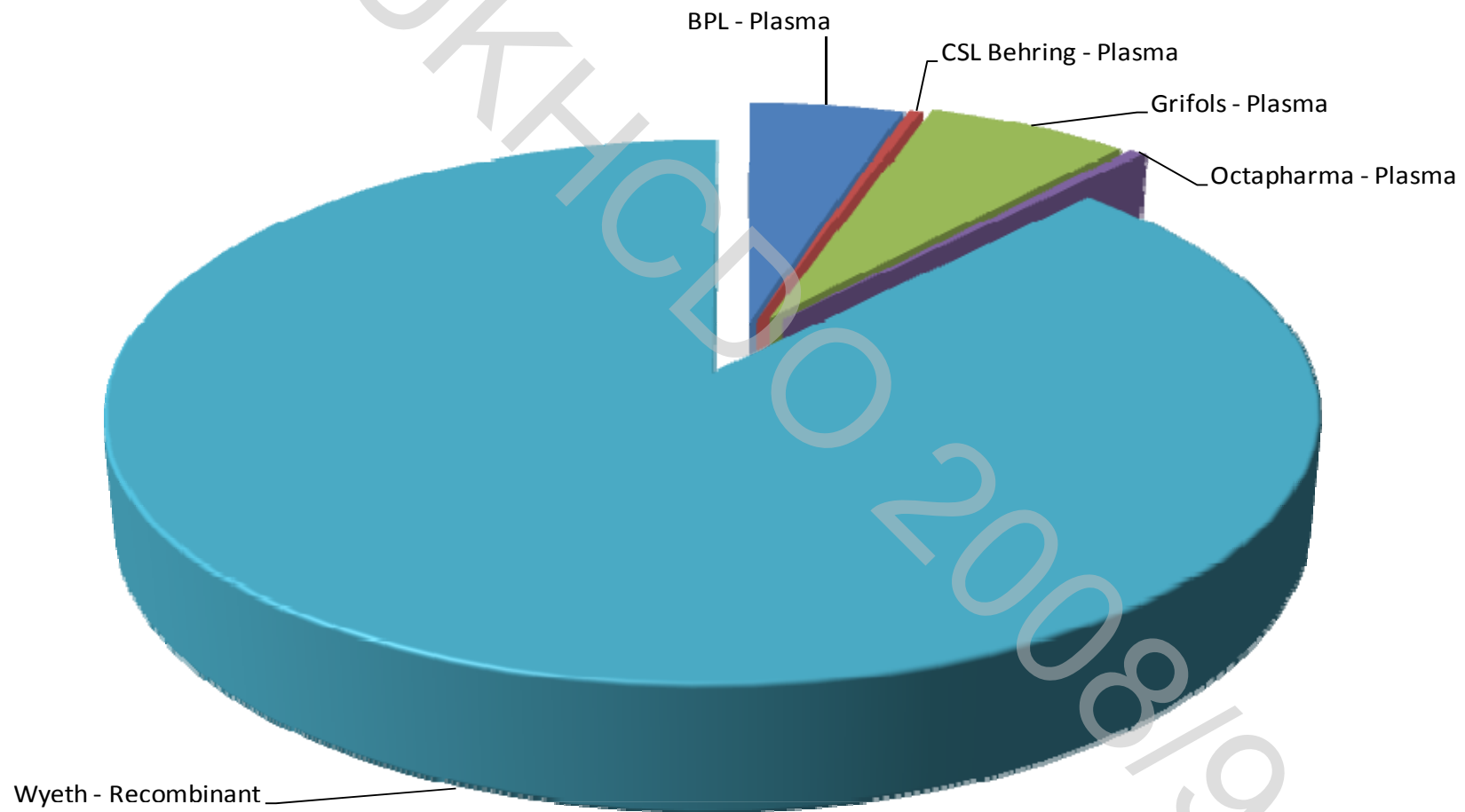
Key	Centre	Patients	Units	Average
1	Birmingham (Queen Elizabeth's)	10	1,826,000	182,600
2	Cambridge	18	3,223,158	179,064
3	Canterbury	6	817,000	136,167
4	Cardiff	9	1,509,000	167,667
5	Glasgow (RI)	15	2,648,656	176,577
6	Hammersmith Hospital, London	3	339,000	113,000
7	Kingston upon Hull	4	698,005	174,501
8	Leicester	5	626,380	125,276
9	Lewisham	6	1,186,914	197,819
10	Manchester (RI)	17	2,651,825	155,990
11	Newcastle uponTyne	11	2,456,000	223,273
12	North Hampshire	4	235,000	58,750
13	Norwich	3	240,000	80,000
14	Oxford	21	5,198,420	247,544
15	Royal Free, London	25	5,841,170	233,647
16	Sheffield (Royal Hallamshire)	10	2,501,745	250,175
17	St George's Hospital, London	4	1,214,680	303,670
18	St Thomas' and Guy's Hospitals, London	20	3,708,730	185,437
19	The Royal London	6	2,022,000	337,000
20	Centres with 1 Patient Meeting Criteria	17	2,725,426	160,319
21	Centres with 2 Patients Meeting Criteria	16	4,636,233	289,765



# Total factor IX units used by UK Haemophilia Centres between 1989 and 2007 and for financial year 2008-2009



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



## Von Willebrand's Sub-Types

VW Sub-types currently in the register	Number
1	2612
1A	8
1B	2
2	108
2A	188
2B	82
2D	1
2M	95
2N	52
3	80
Other	29
Vincenza	2
<b>Subtotal Typed</b>	<b>3259</b>
<b>Type unregistered</b>	<b>6006</b>

## Product usage used in the treatment of von Willebrand's patients

Manufacturer	Product	Total Units
	Cryoprecipitate	10
	DDAVP	116
	Desmopressin-DDAVP (4mcg/ml)	3,027.88
	Desmopressin-Octim nasal spray (150mcg/2.5ml vial)	33,016.75
	Desmopressin-Octim Sub-cut (15mcg/ml)	8,624.26
	Haemocompletten P (Fibrinogen)	7,004
	Platelets	4
<b>Baxter</b>	Advate	98,750
<b>Bayer</b>	Kogenate (Bayer)	8,500
<b>BPL</b>	BPL FVIII 8Y	213,365
	BPL FXI	55,500
	Optivate	194,840
<b>CSL Behring</b>	Beriplex	1,000
	Haemate P	12,704,404
	Helixate Nexgen	5,000
<b>Grifols</b>	Alphanate	2,220,358
	Wilfactin	1,428,451
<b>Novo Nordisk</b>	Novo FVIIa (mg)	190
<b>Octapharma</b>	Wiloctin (Wilate)	322,200

Total Used I.U.  
unless otherwise  
stated

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

Cause of Death	Severity				Total
	Severe	Moderate	Mild	Unknown	
Carcinoma	1	1	4	0	6
Hepatocellular Carcinoma	2	0	0	0	2
Liver Failure	2	2	1	0	5
Cerebral haemorrhage	3	0	3	0	6
Haemorrhage (Misc)	2	1	2	0	5
Ischaemic Heart Disease	2	1	11	0	14
Lymphoproliferative Malignancy	0	0	2	0	2
Motor Neurone Disease	0	0	2	0	2
Senility/Alzheimer's disease	0	0	2	0	2
Venous Thromboembolism	0	0	1	0	1
ARDS	1	0	0	0	1
Intestinal obstruction	0	0	1	0	1
Mixed drug toxicity	0	0	1	0	1
Ruptured Aorta (Peripheral vascular disease)	0	0	1	0	1
Stroke (Unknown)	1	0	0	0	1
Suicide	0	0	1	0	1
Infection (Bacterial)	1	0	4	0	5
COAD	0	0	2	0	2
HIV	0	0	0	0	0
Unknown	5	3	10	1	19
<b>Total</b>	<b>10</b>	<b>2</b>	<b>33</b>	<b>0</b>	<b>45</b>

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

\* 15% of these patients would clear the virus naturally

\*\* When interpreting the number of deaths reported in the group probably infected with hepatitis C it should be recognised that these are deaths from all causes over a period of more than 25 years. No estimate can be given for patients infected with hepatitis C from blood component therapy (plasma and cryoprecipitate). Most of these deaths are not related to hepatitis C. Deaths from hepatitis C typically run at the rate of about 6-8 per year

Diagnosis	Alive at 31/03/2009*	Died by 31/03/2009**	Total
Haemophilia A	1906	1546	3452
Haemophilia B	526	187	713
Females with VIII deficiency	79	18	97
Females with IX deficiency	40	5	45
von Willebrand disease	209	66	275
F.VII deficiency	5		5
F.X deficiency	7	1	8
F.XI Deficiency	4	3	7
F.XII (Hageman) defect	1		1
Prothrombin Deficiency	1		1
Combined von Willebrand's + IX deficiency	1		1
Combined Haemophilia A, Haemophilia B		1	1
Combined Haemophilia A + von Willebrand's	1		1
Combined XI + VIII deficiency	1		1
Combined V+VIII Deficiency	2		2
Combined Haemophilia A + XII deficiency		1	1
Acquired Haemophilia A	2	24	26
Acquired Haemophilia B	1	1	2
Acquired von Willebrands	1	1	2
Platelet defects (misc)	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
Misc.		1	1
Unclassified	9		9
	<b>2,807</b>	<b>1,868</b>	<b>4,675</b>

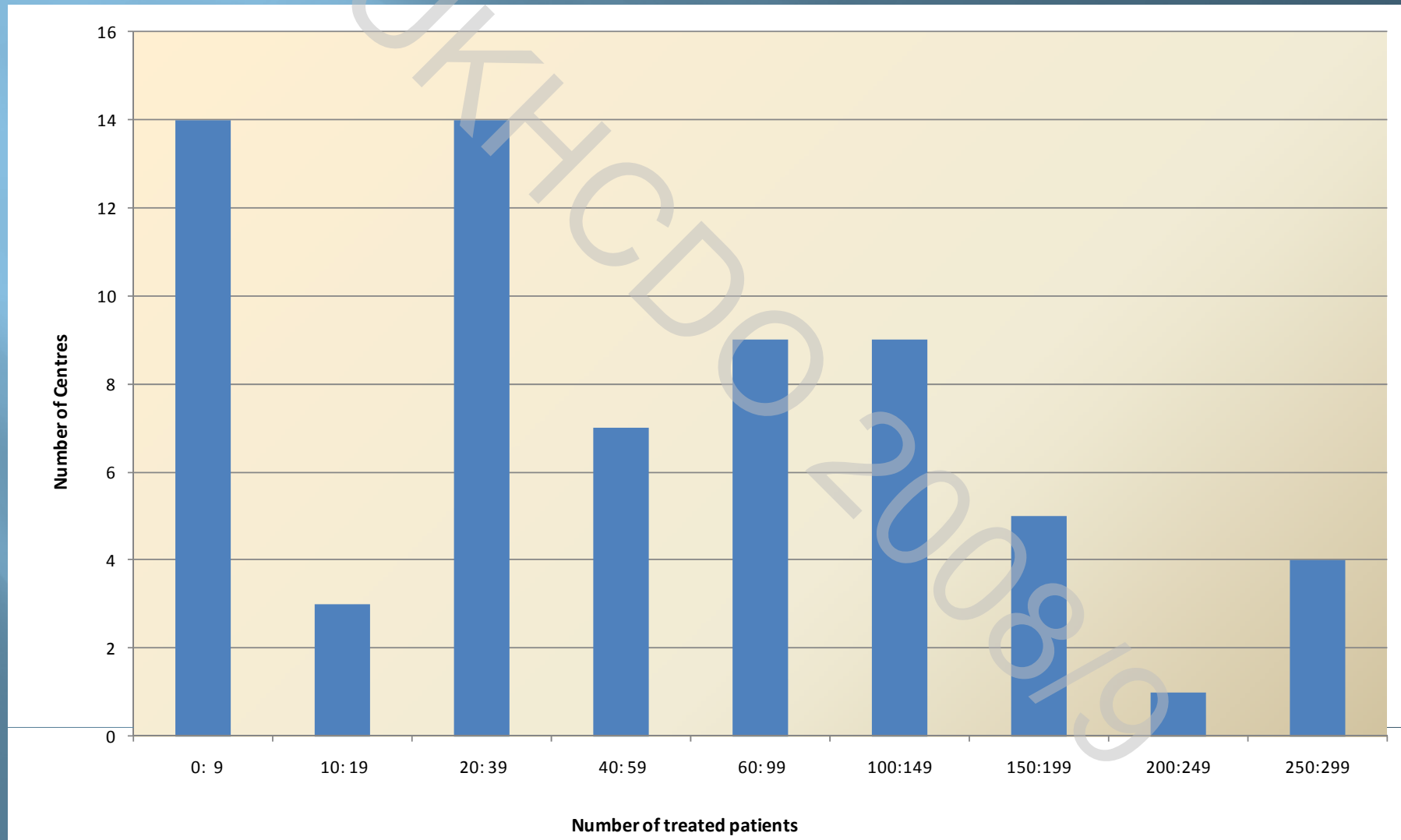
## Annualised UK Deaths from Liver Disease 1969-2009

Year	Hepatoma	Hepatorenal Syndrome	Liver failure	Liver failure associated with cirrhosis and alcoholic liver disease
1969			2	
1970			2	
1972			2	
1973			1	
1974			1	
1975			1	
1979			1	1
1980			1	
1983			0	2
1984			1	2
1985			4	
1986			7	
1987			2	3
1988			1	1
1989			2	3
1990			3	1
1991			9	
1992			9	
1993			14	1
1994			10	3
1995			3	3
1996			8	2
1997			10	
1998			9	
1999			4	
2000			15	
2001			6	
2002	3		4	4
2003	2		1	1
2004	2		3	1
2005	1		1	1
2006	4	1	1	2
2007	2		3	
2008	3		3	
2009	1		5	
<b>Total</b>	<b>18</b>	<b>1</b>	<b>149</b>	<b>31</b>

vCJD - This shows the number of patients, broken down by bleeding disorder diagnosis considered at public health risk of vCJD by virtue of treatment with pooled blood products of UK origin during the period of risk and the number of patients known to have been treated with a batch or batches of concentrate manufactured from a plasma pool containing a donation from a donor who has developed vCJD is also shown

Diagnosis	Number of patients at public health risk	Number of patients known to have received an implicated batch(s)
Haemophilia A	2288	562
Haemophilia B	568	168
Females with VIII deficiency	59	2
Females with IX deficiency	55	3
von Willebrand disease	553	39
F.VII deficiency	35	
F.X deficiency	29	7
F.XI Deficiency	121	
F.XII (Hageman) defect	2	
F.XIII Deficiency	22	
Fibrinogen Deficiency	2	
Combined von Willebrand's + IX deficiency	1	
Combined Haemophilia A + von Willebrand's	1	1
Combined II+VII+IX+X Deficiency	1	
Combined XI + VIII deficiency	1	
Combined XI and XII deficiency	2	
Combined V+VIII Deficiency	7	
Combined Haemophilia A + XII deficiency	2	
Antithrombin III	11	
Combined VII + XI Deficiency	1	
Acquired Haemophilia A	32	5
Acquired Haemophilia B	1	1
Acquired von Willebrands	6	1
Acquired F.XIII Deficiency	1	
Platelet defects (misc)	4	
Severe Platelet Disorders - Other	2	
Haemophilia A with Liver Transplant	10	2
Haemophilia B with Liver Transplant	2	1
Misc.	1	
Unclassified	14	1
Temporary coagulation defect, now normal	2	
<b>Total</b>	<b>3836</b>	<b>793</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

