

## ANNUAL RETURNS FOR 1990

The attached tables and graphs contain the information available from the U.K. Haemophilia Centre Directors Annual Returns. Returns were requested from 112 Centres and have been received from 103 Centres.

By 31.12.90 there were 5,339 haemophilia A patients known to Haemophilia Centres (Table 1). This is an increase of 32 compared with the previous year. Thirty-eight per cent of these patients were severely affected with factor VIII levels <2% average normal. The incidence of antibodies to factor VIII remains as approximately 5% of all haemophilic patients (Table 2).

The total number of patients with Christmas disease now stands at 1055 (Table 3) which is 19 more than in the previous year. Thirty-three per cent of the patients are severely affected. Antibodies to factor IX were reported in 8 cases (Table 2).

The number of patients reported as suffering from von Willebrands disease has risen to 2688 (Table 4) which is 173 more than in 1989. Forty-eight have factor VIII:c levels <2% of average normal and of these 31 required treatment in 1990 and 8 were on home therapy. Forty-four per cent of all von Willebrands patients treated had factor VIII:c levels greater than 30% of normal. Five patients with von Willebrands disease had antibodies to factor VIII (Table 2).

### Treatment of patients

Of the 5,339 haemophiliacs registered 2,388 (45%) received replacement therapy in 1990 (Table 5) and 4,425 (Table 1) were receiving home therapy. The total amount of factor VIII used at Haemophilia Centres in 1990 was 104.9M units an increase of 1,418,000 units over the amount used in the previous year. Haemophiliacs received 100.53M units. (Table 5 and fig. 1). Patients with von Willebrands disease used 3.57M units (Table 5) compared with 2.15M units in 1989. The total amount of NHS factor VIII used rose by 18.75M and the amount of commercial factor VIII fell by 17M compared with 1989.

A total of 4.9M units of human factor VIII was used to treat 131 haemophilia A patients who had factor VIII antibodies (Table 6). In addition 741,000 units of porcine factor VIII, 1.8M units NHS factor IX 2.6M units FEIBA and 174,000 Autoplex were used.

With regard to factor IX, the total amount of factor IX used has decreased since 1989 by 1.9M units to 16.7M units. There has been an increase to a total of 149,000 units in Commercial factor IX used to treat haemophilia B patients. (Table 7 and Fig. 2). 1.8M units of NHS factor IX was used to treat haemophilia A patients who had factor VIII antibodies.

Table 8 gives an analysis of the causes of death in the 87 haemophilic patients, 4 Christmas disease patients and 6 von Willebrands patients

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6th January 1992

WITH COMPLIMENTS

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who died in 1990. AIDS was the commonest cause of death - 51 (53%), with cancer 9 (9%) - the second commonest cause and cerebral haemorrhage - 8 (8%) - the third. Four patients (4%) died from liver disease, 4 (not AIDS cases) from septicaemia and 4 following accidental injuries (2 RTAs, one fall). Further details regarding the cause of death in patients diagnosed as having AIDS are given in Table 9, from which it will be seen that respiratory diseases were the most common cause - 27 (53%). Table 10 shows the HIV status of the patients known to have died in 1990.

An additional table (Table 11) is included this year to show the number of patients with rare blood coagulation defects known to Haemophilia Centre Directors, the number treated and the number on home treatment. Platelet defects form the largest group registered (284), followed by Factor XI deficiency (246) and Factor XII deficiency (124).

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Table 1

Haemophilia A patients known at Haemophilia Centres on 31.12.90, showing the number of patients treated during 1990, with the severity of the coagulation defect and age. Patients known to have died before 1.1.90 have been excluded

Age (years)	Number of Patients Factor VIII Level (%)														
	<2% A.N.			2-10% A.N.			>10% A.N.			Not known			Total		
	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990
<5	135	120	31	83	42	4	44	11	0	1	3	1	263	176	36
5-9	173	151	104	103	61	26	78	13	0	3	2	1	357	227	121
10-14	147	135	121	114	76	34	93	23	3	5	0	0	359	234	158
15-19	164	130	116	128	64	30	113	21	2	11	0	0	416	215	148
20-29	482	373	322	315	132	53	271	46	4	26	1	0	1094	552	379
30-39	409	285	230	269	93	37	221	27	0	27	1	0	926	406	267
40-49	275	189	152	226	63	22	176	25	2	19	1	0	696	278	176
50-59	137	98	71	149	43	10	142	26	2	11	0	0	439	167	83
60-69	66	41	27	110	30	6	136	23	3	11	1	1	323	95	37
70+	39	18	8	72	18	2	93	12	0	8	0	0	212	48	10
Not known	21	1	0	16	0	0	10	1	0	207	4	0	254	6	0
Total	2048	1541	1182	1585	622	224	1377	228	16	329	13	3	5339	2404	1425

Table 2

The number of patients known to Haemophilia Centre Directors on 31.12.90 and the occurrence of Factor VIII and Factor IX antibodies. Patients known to have died before 31.12.90 have been excluded

	Haemophilia A	Haemophilia B	von Willebrands
Cumulative total number of patients known at Centres	5252	1051	2682
Cumulative total number of factor VIII or IX antibodies	283	8	5
% with factor VIII or IX antibodies	5.38	0.76	0.19

Table 3

Haemophilia B patients known at Haemophilia Centres on 31.12.90, showing the number of patients treated during 1990, with the severity of the coagulation defect and age. Patients known to have died before 1.1.90 have been excluded

Age (years)	Number of Patients Factor IX Level (%)														
	<2% A.N.			2-10% A.N.			>10% A.N.			Not known			Total		
	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990
<5	30	22	1	13	9	2	10	1	0	1	0	0	54	32	3
5-9	25	18	14	42	22	8	20	4	0	2	0	0	89	44	22
10-14	32	24	19	36	17	12	25	6	3	1	0	0	94	47	34
15-19	24	17	14	40	23	9	19	5	1	4	0	0	87	45	24
20-29	74	51	47	76	24	11	32	4	0	8	0	0	190	79	58
30-39	55	35	29	62	22	7	44	9	0	5	0	0	166	66	36
40-49	59	35	27	42	10	3	36	6	2	2	0	0	139	51	32
50-59	18	13	10	36	7	5	20	2	0	4	1	1	78	23	16
60-69	15	8	7	24	9	3	15	2	0	1	1	0	55	20	10
70+	8	1	1	25	7	1	14	1	0	6	0	0	53	9	2
Not known	4	0	0	4	0	0	2	0	0	40	0	0	50	0	0
Total	344	224	169	400	150	61	237	40	6	74	2	1	1055	416	237

Table 4

Von Willebrand's disease patients known at Haemophilia Centres on 31.12.90, showing the number of patients treated in 1990, with the Factor VIIIc level and age in 1990. Patients known to have died before 1.1.90 have been excluded

Age (years)	Factor VIIIc Level																	
	<2% A.N.			2-14% A.N.			15-29% A.N.			>30% A.N.			Not known			Total		
	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990	In Reg	Trtd 1990	On HT 1990
<5	1	3	0	5	7	0	0	7	0	83	8	1	0	1	0	89	26	1
5-9	0	4	1	29	9	1	40	7	1	87	14	1	0	1	0	156	35	4
10-14	3	1	0	32	8	1	0	9	1	144	18	1	0	0	0	179	36	3
15-19	0	5	0	36	5	0	65	8	0	137	10	0	0	1	0	238	29	0
20-29	10	3	1	84	20	0	76	22	1	385	26	1	0	2	0	555	73	3
30-39	10	6	2	75	10	4	46	19	1	357	20	2	0	1	0	488	56	9
40-49	7	4	3	55	9	1	39	17	2	323	24	0	0	4	0	424	58	6
50-59	5	2	1	32	5	2	22	8	2	167	10	1	0	1	0	226	26	6
60-69	6	1	0	28	4	1	16	5	0	95	8	0	0	1	0	145	19	1
70+	6	2	0	30	0	0	11	4	0	94	11	0	0	0	0	141	17	0
Not known	0	0	0	0	0	0	0	1	0	0	1	0	47	5	0	47	7	0
Total	48	31	8	406	77	10	315	107	8	1872	150	7	47	17	0	2688	382	33

Table 5  
Factor VIII units used by Haemophilia Centres in 1990

Type of Patient	Total number* of patients treated	Plasma	Cryo.	NHS F.VIII conc.	Commercial F.VIII conc.	Total F.VIII Units
Haemophilia A**	2,388	5,000	53,000	82,469,000	18,003,000	100,530,000
Carriers of Haem. A	31	0	1,000	43,000	0	44,000
von Willebrand's	377	1,000	571,000	2,440,000	559,000	3,571,000
Acquired Haem. A***	18	0	0	743,000	0	743,000
Total	2,814	6,000	625,000	85,695,000	18,562,000	104,888,000
% Total	-	0.01	0.60	81.70	17.70	-

\* Adjusted for duplicates and excluding patients who did not receive blood products

\*\* Including materials used for treatment  
of haemophilia A patients with factor VIII antibodies (See Table 6)

\*\*\* Acquired Haemophilia A patients also received the following materials:-

Porcine F.VIII:	854,000 Units
FEIBA:	1,211,000 FEIBA Units
NHS F.IX:	2,000 Units



Table 6

Materials used by Haemophilia Centres in 1990 to treat 131 Haemophilia A patients who had F.VIII antibodies. In addition, 10 other patients were treated but received no doses

Material	Factor VIII units		Total F.VIII units
	Used at Hosp.	Supplied for HT	
Plasma	240	0	240
Cryoprecipitate	2,000	0	2,000
NHS F.VIII conc.	1,703,000	2,296,000	3,999,000
Commercial Human F.VIII conc.	188,000	713,000	901,000
Total Human F.VIII conc.	1,893,000	3,009,000	4,902,000
Porcine F.VIII	670,000	71,000	741,000
Other Materials:	Units	Units	Units
FEIBA	1,789,000	836,000	2,625,000
AUTOPLEX	174,000	0	174,000
NHS F.IX conc.	508,000	1,331,000	1,839,000

Table 8

Causes of death of 87 Haemophilia A, 4 Haemophilia B and 6 von Willebrand's disease patients in 1990, showing severity of coagulation defect and age at death

Cause of death	F.VIII/IX level %	Age at Death (years) and Number of Deaths*									TOTAL
		<5	5-9	10-19	20-29	30-39	40-49	50-59	60-69	70+	
AIDS**	<2	-	1	5(1)	8(1)	15(3)	4(1)	6(2)	2	-	
	2-10	-	-	1	1	1	2	1	-	-	5(2)
	>10	-	-	-	-	-	1	1	-	-	
	N/K	-	1	-	-	-	-	-	-	1	
Cerebral Haemorrhage/Stroke	<2	-	-	-	-	1	-	3(1)	-	1(1)	
	2-10	-	-	-	-	-	-	-	1	1	8(2)
	>10	-	-	-	-	-	-	-	1	-	
Ischaemic Heart Disease	<2	-	-	-	-	-	-	-	-	1(1)	
	2-10	-	-	-	-	-	-	-	1	-	3(1)
	>10	-	-	-	-	-	-	-	1	-	
Cancer	<2	-	-	-	-	-	1	-	1	1(1)	
	2-10	-	-	-	1	-	-	1	1	2	9(1)
	>10	-	-	-	-	-	-	-	-	1	
Hodgkins Disease/ Lymphoma	<2	-	-	-	1	-	-	-	-	-	2
	2-10	-	-	1	-	-	-	-	-	-	
Acute Myeloid Leukaemia	2-10	-	-	-	-	-	-	-	-	1	1
Liver disease	<2	-	-	-	-	-	1	2(1)	-	1	4(1)
Pneumonia (Not PCP)	<2	-	-	-	-	-	-	-	1(1)	-	1(1)
Septicaemia	<2	-	-	-	-	-	1	1	-	-	4(1)
	2-10	-	-	-	-	-	1(1)	-	1	-	
Extensive abdominal haemorrhage	<2	-	-	-	-	-	-	-	1(1)	-	1(1)
G.I. Bleed	<2	-	-	-	-	-	-	-	-	1(1)	1(1)
Pulmonary Embolism	>10	-	-	-	-	-	-	-	1	-	1
Chronic respiratory disease	2-10	-	-	-	-	-	-	-	1	-	2
	>10	-	-	-	-	-	-	-	-	1	
Post-operative infection	>10	-	-	-	-	-	-	1	-	-	1
Fibrinous pericarditis	<2	-	-	-	-	-	-	1(1)	-	-	1(1)

Table 7

Factor IX units used by Haemophilia Centres in 1990

Type of patient	Total number of patients*	Plasma	NHS IX	Commercial IX	Total units
Haemophilia B**	416	2,000	15,208,000	149,000	15,359,000
Carriers of Haemophilia B	13	0	91,000	0	91,000
Haem. A patients with F.VIII antibodies	22	0	1,839,000	0	1,839,000
Total	448	2,000	17,138,000	149,000	17,288,000
% Total	-	0.01	99.10	0.88	-

\* Adjusted for duplicates and excluding patients who did not receive blood products

\*\* Including 5 Haemophilia B patients with Factor IX antibodies

1 Haemophilia B patient with F.IX antibodies also received 24,500 units of FEIBA and  
1 only received 11,520 units of Autoplex

Table 8 (cont.)

Cause of death	F.VIII/IX level %	Age at Death (years) and Number of Deaths*									
		<5	5-9	10-19	20-29	30-39	40-49	50-59	60-69	70+	TOTAL
Road Traffic Accident	<2 2-10	-	-	-	2	-	-	-	-	-	3
Accidental fall	>10	-	-	-	-	-	-	-	-	1	1
Overdose	<2	-	-	-	-	1	-	-	-	-	1
Alzheimers Disease	>10	-	-	-	-	-	-	-	1	-	1
Not known	<2	-	-	-	-	-	-	-	-	1	1
TOTAL		-	2	8(1)	13(1)	18(3)	11(2)	17(5)	14(2)	14(4)	97(17)

\* Figs. in brackets ( ) = Number of patients who had F.VIII antibodies

\*\* See Table 9 for further details

There were also 6 other deaths during 1990:

4 patients with Acquired Haemophilia (G.I. bleed; cerebral haemorrhage; Haemorrhage (unspecified); ilio-psoas bleed).

1 patient with Acquired von willebrand's Disease (G.I. bleed)

1 patient with Factor XI (PTA) deficiency (Ischaemic heart disease)

Table 9  
Cause of Death During 1990 in Patients Diagnosed as having AIDS

Clinical Information/Autopsy Details	Number of Patients
<u>Respiratory Diseases:</u>	
PCP	8
PCP, respiratory failure, hepatic failure	1
Probable PCP, cerebral atrophy, encephalitis	1
Cryptosporidium, PCP	1
? PCP, mycobacterium avium infection	1
Lymphoma, PCP	1
Pneumonia/ bronchopneumonia	6
Bronchopneumonia, lymphoma, PCP, mycobacterium avium	1
Bronchopneumonia, cryptosporidial infection	1
Pneumonia, severe wasting	1
Pseudomonas pneumonia	1
Pneumonia, pneumothorax	1
Fil + pneumonia	1
Cerebral toxoplasmosis, haemorrhagic, bronchopneumonia	1
Bronchopneumonia and dementia	1
Respiratory failure secondary to pneumothorax	1
Respiratory failure	1
<u>Cerebral Infections</u>	
Encephalitis	1
HIV-related encephalopathy	1
Cerebral AIDS/dementia	1
<u>Other Infections</u>	
CMV	1
Septicaemia	1
Disseminated atypical mycobacterial infection, bilateral pneumothorax, disseminated CMV	1
<u>Other Conditions</u>	
AIDS Wasting Syndrome	2
Lymphoma	2
G.I. Haemorrhage, encephalopathy	1
Cerebral haemorrhage	3
Intraperitoneal haemorrhage	1
Liver disease	2
Myocardial infarction	1
No details	4
TOTAL	51

**Table 10**  
HIV Status of Haemophilia A, Haemophilia B and von Willebrand's disease patients known to have died during 1990

Cause of Death	Number of Patients			
	HIV Status		Not Tested	Total
	Positive	Negative		
AIDS	51	-	-	51
Cerebral haemorrhage/stroke	3	4	1	8
Ischaemic heart disease	1	1	1	3
Cancer	4	5	-	9
Hodgkins Disease/Lymphoma	-	-	2	2
Acute myeloid leukaemia	-	1	-	1
Liver disease	1	3	-	4
Pneumonia (not PCP)	1	-	-	1
Septicaemia	3	1	-	4
Extensive abdominal haemorrhage	-	1	-	1
Gastro-intestinal bleed	-	1	-	1
Pulmonary embolism	-	1	-	1
Chronic respiratory disease	-	-	2	2
Post-operative infection	-	-	1	1
Fibrinous pericarditis	1	-	-	1
Overdose	-	1	-	1
Road Traffic Accident	2	1	-	3
Accidental fall	-	-	1	1
Alzheimers Disease	-	-	1	1
Not known	-	1	-	1
TOTAL	67	21	9	97

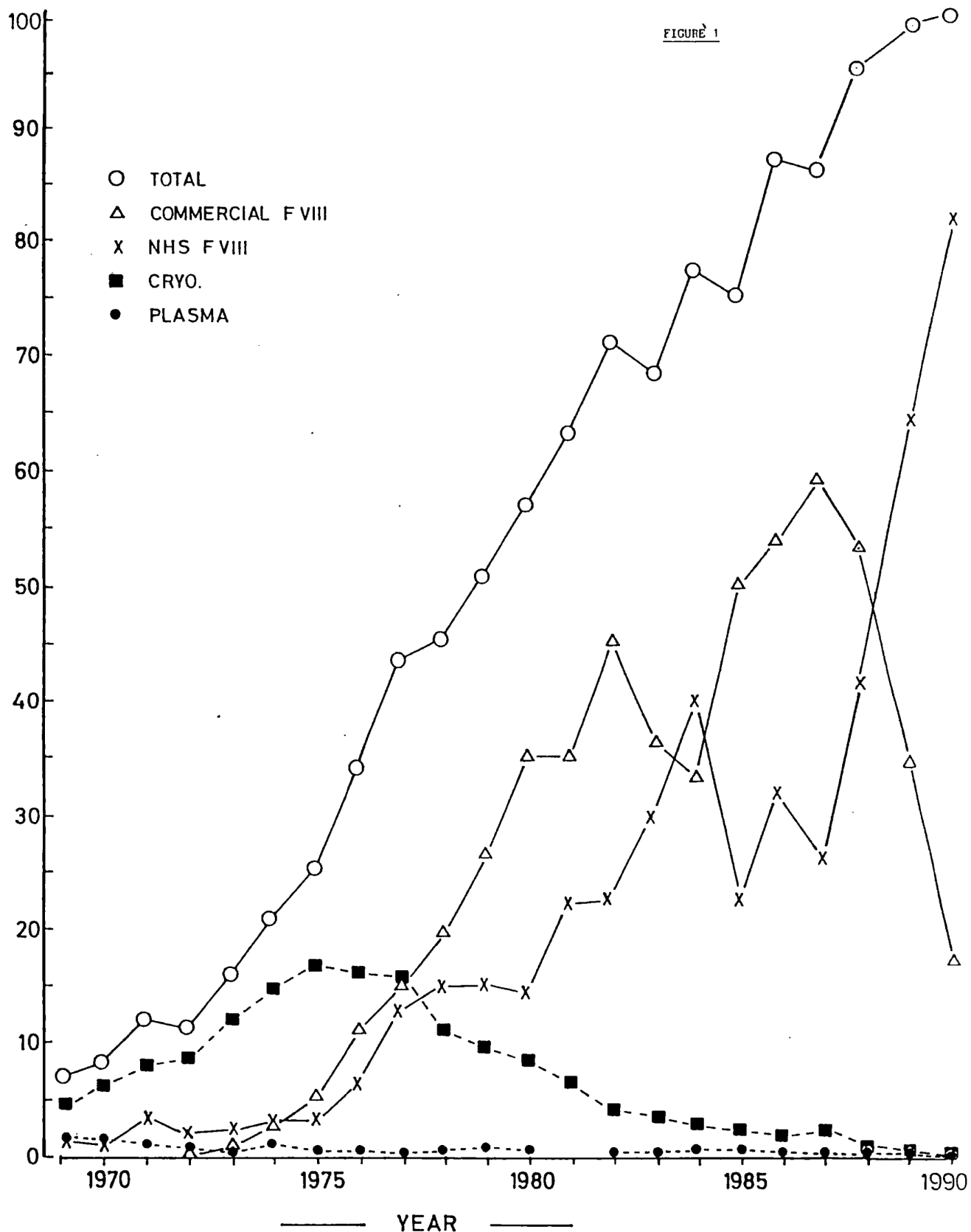
Table 11

Number of Patients with rare congenital blood coagulation defects known to UK Haemophilia Centres, showing the number treated during 1990 and the number on home treatment

<u>Coagulation Defect</u>	<u>Number of Patients</u>		
	In Register in 1990	Treated in 1990	On HT in 1990
Congenital Fibrinogen Deficiency	35	5	-
Congenital Prothrombin Deficiency	12	1	-
Factor V Deficiency	21	3	-
Factor VII Deficiency	59	7	-
Factor X Deficiency	48	10	2
Factor XI (PTA) Deficiency	246	14	1
Factor XII (Hageman) Defect	124	1	-
Factor XIII (FSF) Deficiency	21	17	1
Combined XI + XII Deficiency	3	-	-
Combined II + VII + IX + X Deficiency	7	-	-
Combined V + VIII Deficiency	17	2	-
Combined XI + VIII Deficiency	6	-	-
Combined VIII + IX Deficiency	1	-	-
Combined IX + XI Deficiency	1	-	-
Antithrombin III Defect	35	1	-
Platelet Defects	284	14	-
Fletcher Factor	6	-	-
TOTAL	926	75	4

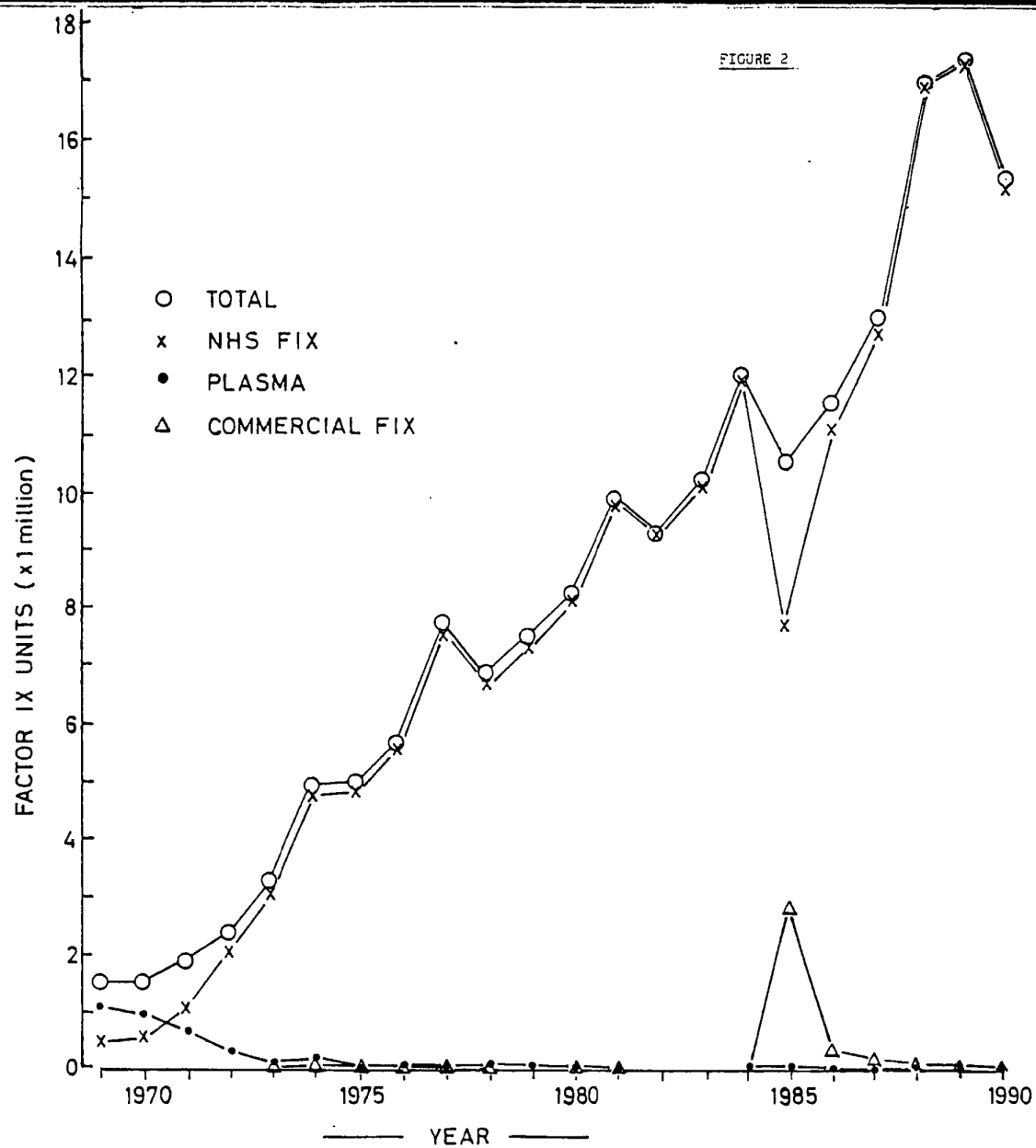
FIGURE 1

FACTOR VIII UNITS (x1 million)



AMOUNT OF BLOOD PRODUCTS (F.VIII UNITS) USED TO TREAT HAEMOPHILIA A PATIENTS IN THE UK





AMOUNT OF BLOOD PRODUCTS (F.IX UNITS) USED TO TREAT HAEMOPHILIA B PATIENTS IN THE UK