

Annual Returns for 1982

The attached tables and graphs outline the information available from the U.K. Haemophilia Centre Directors Annual Returns received by mid-July 1983. Please note that this is an incomplete report. We have prepared it in July to allow time for it to be circulated before the A.G.M., but we are still waiting for the returns to be completed by several Centres.

Number of patients

By 31.12.82 there were 4,592 haemophilia A patients known to Haemophilia Centres (Table 1). Forty three per cent of those patients were severely affected with factor VIII levels <2% average normal. The incidence of antibodies to factor VIII is still approximately 6% of all patients (Table 2). During 1982 there was an increase in the number of patients with newly detected antibodies; 25 new antibody cases were reported compared with 18, 12 and 9 in 1979, 1980 and 1981 respectively (Fig. 1). As in previous years there are still a large number of patients whose age or level of factor VIII are not known. In 172 patients neither age nor factor VIII level is known; 13 of those patients received replacement therapy during 1982 (Table 1).

The number of patients with Christmas disease known at Haemophilia Centres now stands at 823 of whom 281 (34%) are severely affected (Table 3). Antibodies to factor IX were reported in 8 cases (Table 2). As in the case of haemophiliacs information concerning factor level and age was not available in a number of patients: in 34 cases neither age nor factor IX level was known (Table 3).

Treatment of patients

Of the 4,592 registered haemophiliacs, 2,211 (48%) received replacement therapy during 1982 (Table 1), and 1,101 were receiving home treatment; 50% of severely affected patients are now on home treatment. It is interesting that nearly 10% of mildly affected haemophiliacs (factor VIII > 10%) are receiving home treatment.

The total amount of factor VIII used to treat haemophiliac patients is now 71M units and rises to nearly 74M units if the amount of factor VIII used in the management of von Willebrand's disease is included (Table 4). The latter figure is an increase of 12% over the total amount used in 1981. The continued increase in the amount of factor VIII used to treat haemophiliac patients is shown in Fig. 2, from which it can be seen that the amount of commercial concentrates continues to rise. The amount of NHS concentrates used has shown little change since 1981 and the amount of Cryoprecipitate used continues to fall. The average amount of factor VIII used for the treatment of haemophilia in 1982 was nearly 33,000 units per patient. Approximately half of the factor VIII used in the management of haemophiliacs was used as home treatment (Table 5).

1686(a)

46/310

Nearly 6M units of factor VIII were used in the treatment of 144 patients who had factor VIII antibodies, the majority of this material was commercial factor VIII (Table 6). In addition nearly 1.2M units of porcine factor VIII were used, 1.39M units of NHS factor IX, 1.56M units of FEIBA and 0.5M units of Autoplex. The record of the amount of Autoplex used is incomplete as some Centres participating in a double-blind clinical trial were unable to say how much they had used.

With regard to the treatment of patients with Christmas disease the amount of factor IX used fell during 1982 (Fig. 3 and Table 7), but the overall usage of factor IX has increased because of the large amounts used in the management of haemophilia A patients who have factor VIII antibodies (Tables 6 and 7). Forty-four per cent of the total amount of material used to treat Christmas disease patients was used for home therapy (Table 9). The amount of factor IX used for the management of patients with factor IX antibodies is shown in Table 8.

Table 10 shows the type and amount of material used to treat von.Willebrand's disease patients. Twenty-six per cent of the material was used for home treatment.

Table 11 gives an analysis of the cause of death in the 21 haemophilic patients and 2 Christmas disease patients who died. Nine (39%) deaths were due to cerebral haemorrhage and 6 of those were in patients more than 50 years of age. Other types of haemorrhage accounted for a further 5 deaths.

C.R. Rizza  
Rosemary J.D. Spooner

Oxford Haemophilia Centre

31.7.83

46/311

Table 1

Haemophilia A patients known at Haemophilia Centres in 1982, showing the number of patients treated during 1982, with the severity of the coagulation defect and age. Patients known to have died before 1.1.82 have been excluded

46/312

Age (Years)	Number of Patients Factor VIII level (%)														
	<2% A.N.			2-10% A.N.			>10% A.N.			Not known			Total		
	In Reg	Trtd	On HT	In Reg	Trtd	On HT	In Reg	Trtd	On HT	In Reg	Trtd	On HT	In Reg	Trtd	On HT
<5	116	84	5	59	34	0	28	10	0	5	3	0	208	131	5
5-9	143	114	62	104	65	12	63	19	1	7	0	0	317	198	75
10-19	454	380	307	257	120	43	191	46	1	37	4	0	939	550	351
20-29	452	349	270	266	107	24	173	31	0	30	3	0	921	490	294
30-39	347	254	183	214	66	12	139	28	2	31	6	0	731	354	197
40-49	210	148	100	165	49	8	114	18	1	22	1	0	511	216	109
50-59	121	81	44	116	32	8	101	26	3	12	3	0	350	142	55
60-69	65	38	11	93	28	1	70	13	1	17	3	0	245	82	13
70+	30	12	2	57	11	0	52	7	0	8	0	0	147	30	2
Not known	22	2	1	21	3	0	8	0	0	172	13	0	223	18	1
Total	1960	1462	985	1352	515	108	939	198	9	341	36	0	4592	2211	1102

Table 2

The number of patients known to Haemophilia Centre Directors on 31.12.82 and the occurrence of Factor VIII and Factor IX antibodies. Patients known to have died before 31.12.82 have been excluded

	Haemophilia A	Haemophilia B	Von Willebrands
Cumulative total number of patients known at Centres	4,571	821	1,178
Cumulative total number with factor VIII or IX antibodies	266	8	4
% with factor VIII or IX antibodies	5.82	0.97	0.34

Table 3

Haemophilia B patients known at Haemophilia Centres in 1982, showing the number of patients treated during 1982, with the severity of the coagulation defect and age. Patients known to have died before 1.1.82 have been excluded

46/314

Age (Years)	Number of Patients Factor IX level (%)														
	<2% A.N.			2-10% A.N.			>10% A.N.			Not known			Total		
	In Reg	Trtd	On HT	In Reg	Trtd	On HT	In Reg	Trtd	On HT	In Reg	Trtd	On HT	In Reg	Trtd	On HT
<5	19	16	2	16	10	1	8	3	0	1	1	0	44	30	3
5-9	23	18	11	22	8	4	11	3	0	2	0	0	58	29	15
10-19	69	55	45	70	31	12	32	7	1	7	0	0	178	93	58
20-29	58	40	27	63	27	9	37	14	1	6	1	0	164	82	37
30-39	58	40	27	46	12	2	29	6	1	7	1	0	140	59	30
40-49	21	18	10	27	9	2	13	2	0	9	3	0	70	32	12
50-59	16	11	4	25	9	1	12	4	0	3	2	2	56	26	7
60-69	9	3	1	20	6	0	9	1	0	7	2	0	45	12	1
70+	4	2	0	12	2	1	6	0	0	2	0	0	24	4	1
Not known	4	0	0	3	0	0	3	0	0	34	4	0	44	4	0
Total	281	203	127	304	114	32	160	40	3	78	14	2	823	371	164

Table 4

Factor VIII Units Used by Haemophilia Centres in 1982

46/315

Type of Patient	Total Number* of Patients Treated	Plasma	Cryo.	NHS F.VIII Conc.	Commercial F.VIII Conc.	Total F.VIII Units	% Total F.VIII Units	Average Amount Used per Patient
Haemophilia A**	2,186 <sup>6</sup>	1,000	3,851,000	22,475,000	44,978,000	71,305,000	96.71	32,619
Carriers of Haem. A	34 <sup>66</sup>	Nil	45,000	37,000	16,000	98,000	0.13	2,882
Von Willebrand's	258 <sup>666</sup>	45,000	1,254,000	380,000	650,000	2,329,000	3.16	9,027
TOTAL	2,478	46,000	5,150,000	22,892,000	45,644,000	73,732,000	100	29,755
%Total	-	0.06	6.99	31.04	61.91	100	-	-

\* Adjusted for duplications

\*\* Additional materials used for Haemophilia A patients with F.VIII antibodies:-

NHS F.IX Concentrate	1,388,000 units
Porcine F.VIII Concentrate	1,183,000 units
FEIBA	1,563,000 units
AUTOPLEX	501,000 units + ?

<sup>6</sup>Plus 26 who received no doses.

<sup>66</sup>Plus 3 who received no doses.

<sup>666</sup>Plus 20 who received no doses.

Table 5

Materials used for treatment of Haemophilia A patients during 1982 and the amount used for Home Treatment

46/316

Material	Total F.VIII Units used for all Haem. A Patients	F.VIII units used for Home Treatment only	% used for Home Treatment
Plasma	1,000	Nil	-
Cryoprecipitate	3,851,000	834,000	21.66
NHS F.VIII Conc.	22,475,000	13,109,000	58.33
Commercial F.VIII Conc.	44,978,000	24,942,000	55.45
Total Units	71,305,000	38,885,000	54.53
Number of patients treated	2,186	1,102	50.41
Average amount used per patient	32,619	35,286	-

In addition, NHS F.IX, PORCINE F.VIII, FEIBA and AUTOPLEX were supplied for HT to Haemophilia A patients with F.VIII antibodies (See Table 3).

Table 6

46/317

Materials used by Haemophilia Centres in 1982 to treat 144 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	-	-	-
Cryo.	71,000	16,000	87,000
NHS F.VIII Conc.	460,000	303,000	763,000
Commercial Human F.VIII Conc.	3,409,000	1,592,000	5,001,000
Total Human F.VIII Conc.	3,940,000	1,911,000	5,851,000
Porcine F.VIII	1,180,000	3,000	1,183,000
Other Materials:	Units	Units	Total Units
FEIBA	1,521,000	42,000	1,563,000
AUTOPLEX	474,000	27,000	501,000
NHS F.IX Conc.	426,000	962,000	1,388,000



Table 7

Factor IX Units Used by Haemophilia Centres in 1982

46/318

Type of Patient	Total number* of Patients Treated	Plasma	NHS F.IX	Total Units	% Total F.IX Units	Average Amount Used per Patient
Haemophilia B	370	Nil	9,252,000	9,252,000	86.72	25,005
Carriers of Haem. B	9	5,000	24,000	29,000	0.27	3,222
Haem. A Patients with F.VIII Antibodies	11	-	1,388,000	1,388,000	13.01	126,182
TOTAL	390	5,000	10,664,000	10,669,000	100	27,356
% Total		0.05	99.95	100	-	

\* Adjusted for duplicates

Table 8

Material used by Haemophilia Centres in 1982 to treat 3  
Haemophilia B (Christmas disease) patients who had F.IX  
antibodies

Material	Factor IX Units		Total F.IX Units
	Used at Hosp.	Supplied for HT	
NHS F.IX Conc.	77,000	35,000	112,000

46/319

Table 9

Materials used for treatment of Haemophilia B patients during 1982 and the amount used  
for Home Treatment

46/320

Material	Total F.IX units used for all Haemophilia B patients	F.IX units used for Home treatment only	% used for Home treatment
NHS F.IX Concentrate	9,252,000	5,400,000	58.37
Number of patients treated	370	164	44.32
Average amount used per patient	25,005	32,927	-

46/321

Table 10

Materials used for treatment of von Willebrand's disease patients during 1982 and the amount used for Home Treatment

Material	Total F.VIII units used for all von Willebrands patients	F.VIII units used for Home Treatment only	% used for Home Treatment
Plasma	45,000	Nil	-
Cryoprecipitate	1,254,000	262,000	20.89
NHS F.VIII Concentrate	380,000	162,000	42.63
Commercial F.VIII Conc.	650,000	182,000	28.00
Total Units	2,329,000	606,000	26.02
Number of patients treated	258	14	5.43
Average amount used per patient	9,027	43,286	-

Table 11

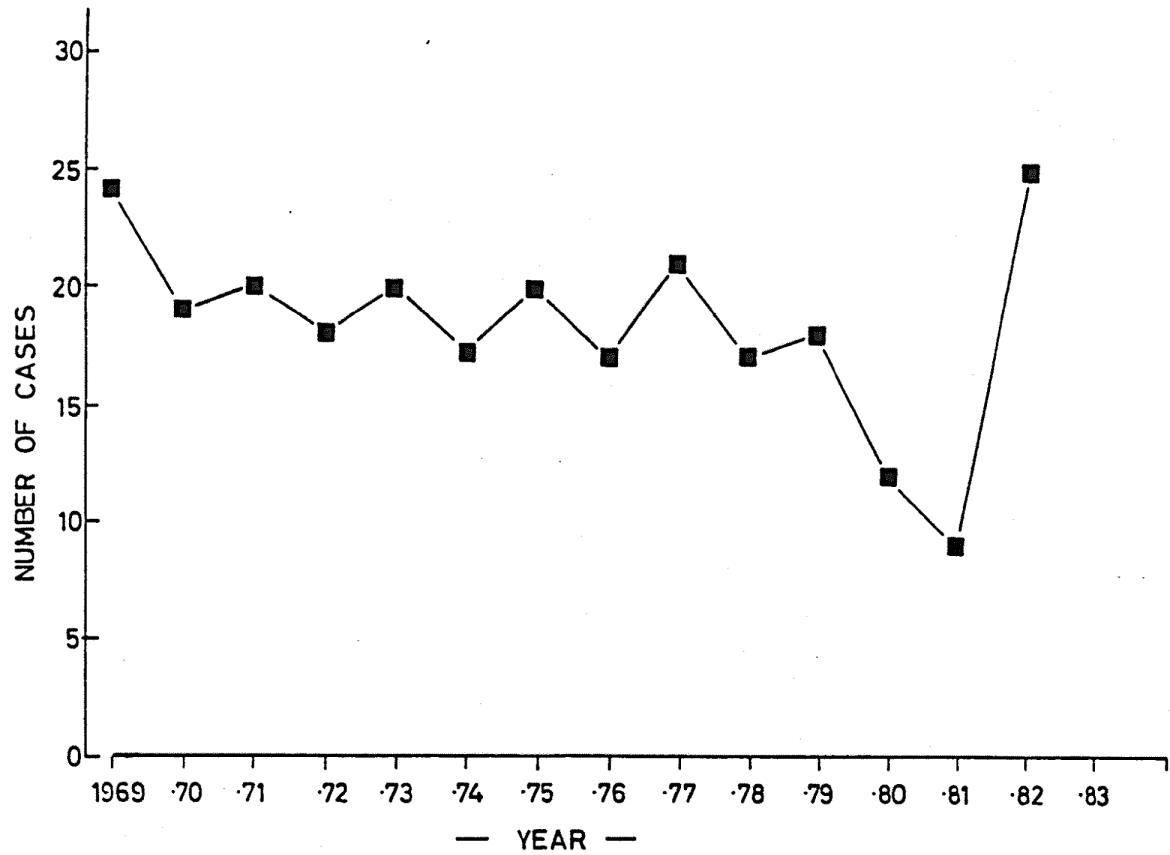
Cause of death in 21 Haemophilia A and 2 Haemophilia B patients during 1982, showing severity of coagulation defect and age at death

46/322

Cause of Death	Factor VIII/IX level %	Number of Deaths*								
		Age at Death (years)								TOTAL
		<10	10-19	20-29	30-39	40-49	50-59	60-69	70+	
Cerebral haemorrhage	<2 2-10 >10	1 - -	- - -	1 - -	1(1) - -	- - -	- - 1	2(1) 2 -	- 1 -	9(2)
Intra-Abdominal haem.	<2	-	1(1)	-	-	-	-	-	-	1(1)
Ruptured spleen	<2	-	1	-	-	-	-	-	-	1
Haemorrhage from bilateral renal artery aneurysms	2-10	-	-	-	-	1	-	-	-	1
G.I. bleed	<2	-	-	-	-	1	-	-	-	1
Throat haemorrhage	2-10	-	-	-	-	-	1	-	-	1
Pneumonia	2-10	-	-	-	-	-	-	1	-	1
Septicaemia	<2	-	-	1(1)	-	-	1	-	-	2(1)
Accident	N/K	-	-	-	-	1	-	-	-	1
Cancer	2-10 >10	- -	- -	- -	- -	- -	- 1	1 -	- -	2
Intestinal obstruction	<2 2-10	- -	- -	- 1	- -	- -	- -	- -	1 -	2
Cardiac failure	<2	-	-	1	-	-	-	-	-	1
TOTAL		1	2(1)	4(1)	1(1)	3	4	6(1)	2	23(4)

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

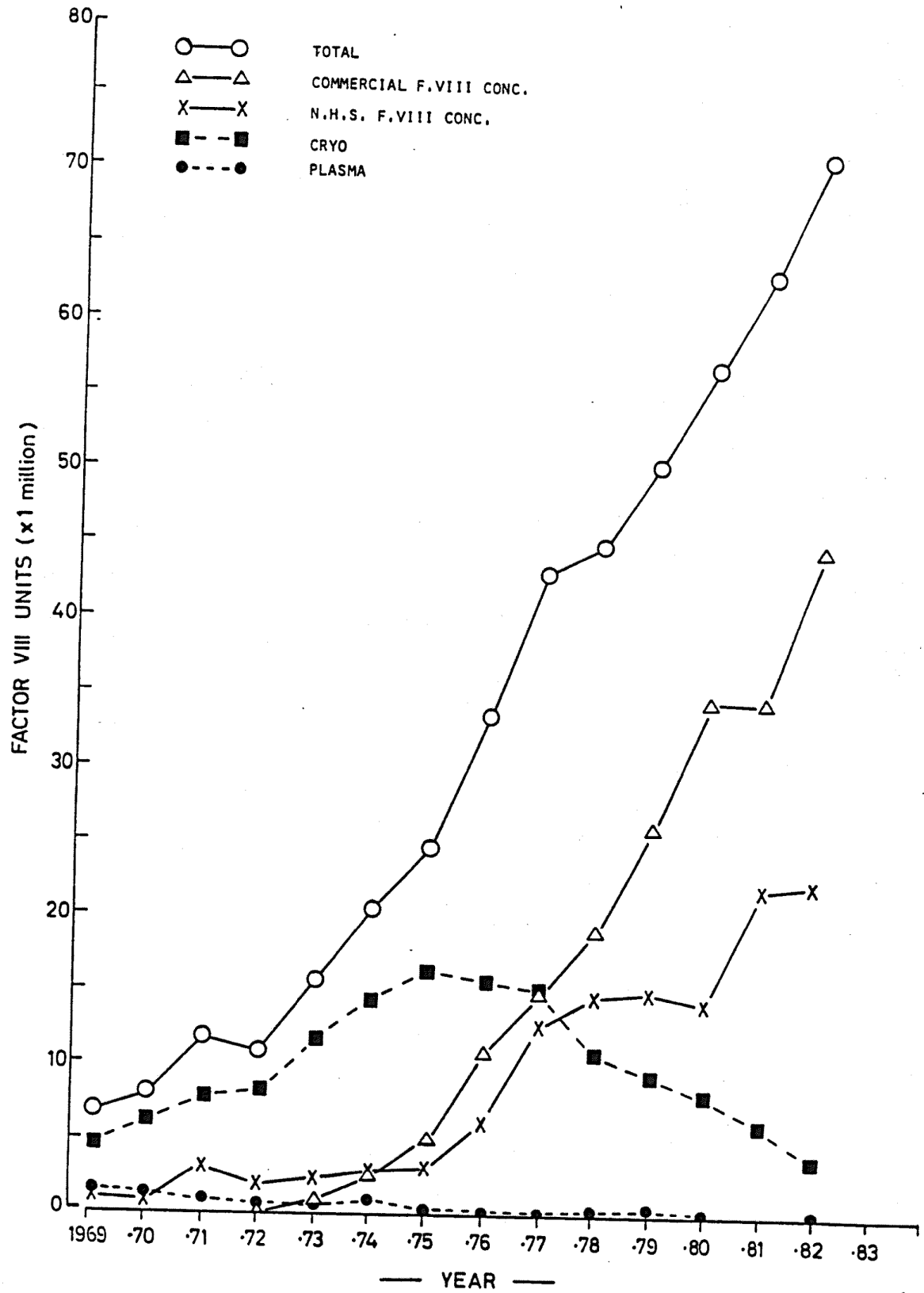
FIG. 1.



Number of new cases of F.VIII Antibodies (inhibitors) in Haemophilia patients detected per year

46/32.

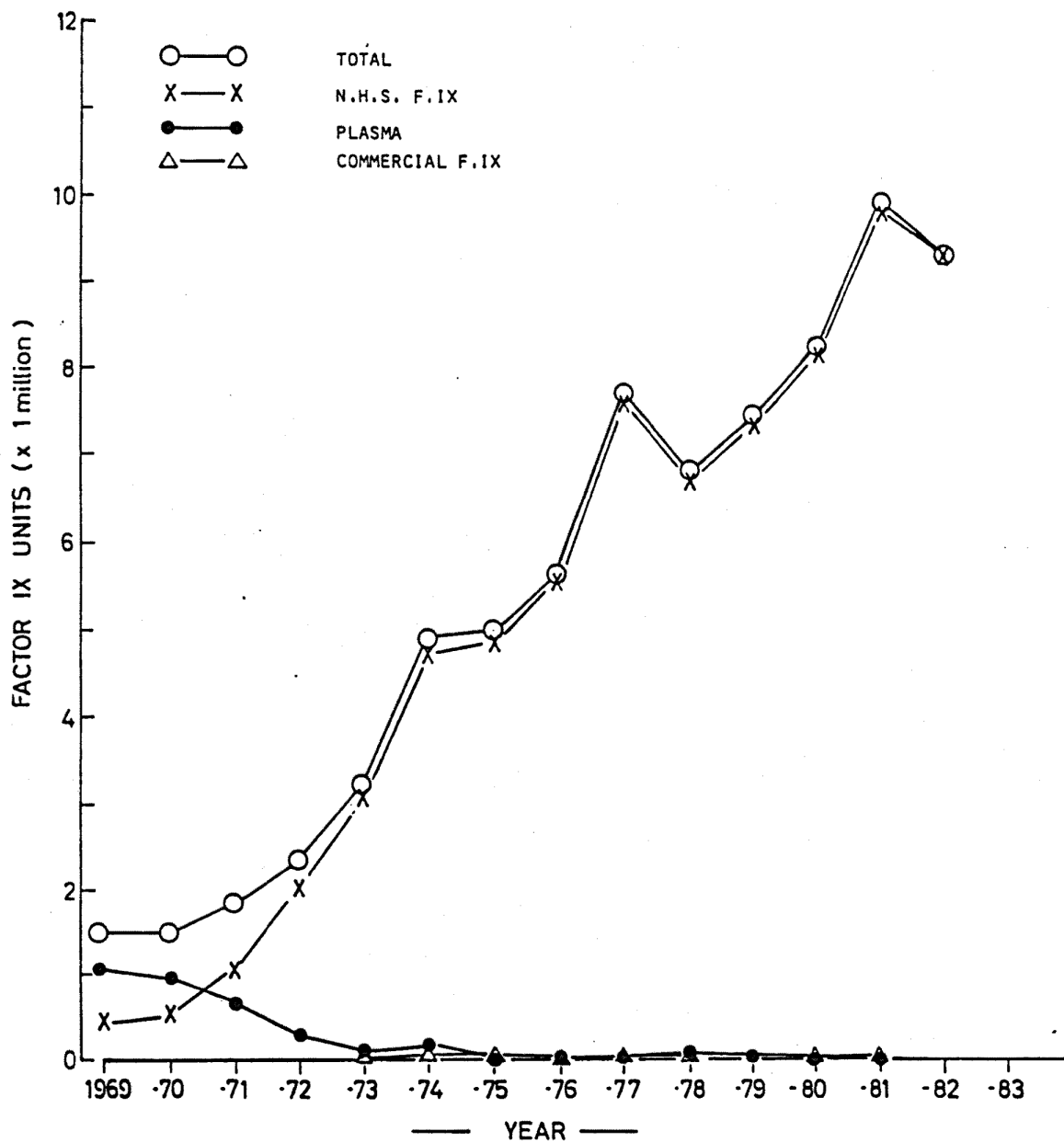
FIG. 2.



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

46/32

FIG. 3.



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

46/32-