CARE OF HAEMOPHILIACS IN THE LONDON AREA

This memorandum is intended as a basis for discussion of the need to improve existing facilities for the care of patients with haemophilia and related disorders in and around London. With the increasing availability of cryoprecipitate, a much more active approach can and should be taken to the treatment of minor bleeding episodes. The concept of an adequate therapeutic service has therefore altered radically during the last few years, and the provision of such a service now demands much greater resources of both staff and therapeutic materials than formerly. We have attempted to set out the present requirements in relation to the number of treatments given annually by a Centre.

Functions of a Haemophilia Centre in the London Area

The functions of a Haemophilia Centre are to provide a diagnostic service in the field of congenital bleeding disorders and to accept the care of patients found to be suffering from haemophilia and allied conditions, including the provision of treatment at short notice and of an advisory service to patients and general practitioners (HM(68)8). The existence of these facilities inevitably attracts a good deal of additional work of a diagnostic and advisory nature within the wider field of haemostatic disorders in general.

To ensure maximal benefit to the patient, with early return to work or school, and minimal residual disability, treatment should be given as early as possible after the onset of a haemophilic bleed. Although many of our haemophilic patients live outside the London

area (only half of those seen in recent years at Great Ormond Street and St. Thomas's Hospital, for example, live within the London postal districts), the transport network is such that most are able to attend early in the development of a bleed, and all are encouraged to do so. The major therapeutic function of a Centre in London is therefore to provide a "demand" treatment service by day and night. Advice should also be available on social and educational problems. The larger Centres should provide antihaemophilic cover for emergency and certain elective surgery (including dental extractions); though some major elective surgery may still have to be referred to Oxford according to the overall policy already laid down. It seems to us also appropriate for a large Haemophilia Centre to be able to make routine assays of factor-VIII potency of cryoprecipitate made at its regional Blood Transfusion Centre so as to provide the Centre with regular feed-back on its material.

Our three Centres already attempt to fulfil all these functions, but with our present resources we are unable to do so to our satisfaction. The proper care of patients demands a great deal more detailed monitoring of therapeutic materials, and of responses to therapy, than we are at present able to carry out, even by diverting research staff onto such work.

Measure of the Work Load and Therapeutic Requirements at a Haemophilia Centre

The total number of patients under care at a Centre is not a very precise measure of the work load, because the volume of work will depend on the proportion of cases who are severely affected and on the

proportion who live near enough to be easily treated as outpatients (in London this proportion should be high). The quantity of therapeutic materials required will depend, again, on the proportion of severely affected patients and on their age distribution, since dose is related to body weight.

We suggest that the most reliable measure of work load is the average number of treatments given per annum. For this purpose, we define a "treatment" as a single dose of therapeutic material (plasma, cryoprecipitate, etc.) of whatever size is appropriate; a single bleeding episode may require one or many such treatments. The work may be broken down into treatments for outpatients and for inpatients, (as shown for our three hospitals in Table I), or into treatments for bleeding episodes and elective treatments to prevent bleeding after surgery and dental extractions, as shown in Figure 1 for St. Thomas's Hospital. An indication of the distribution of different bleeding disorders in given in Table II.

TABLE I

Current work load at the three Haemophilia Centres of the Hospital for Sick Children, Great Ormond Street, (G.O.S.), Royal Free Hospital, Lawn Road Branch (R.F.H.) and St. Thomas's Hospital (S.T.H.). January to October 1969, inclusive.

	R.F.H.	S.T.H.	G.O.S.
No. of patients under supervision (approx.)	160*	180	90*
No. of patients treated	78	58	61
No. of treatments	1051	931	894
Inpatients	199	422	396
Outpatients	852	509	498
Average no. of treatments per patient treated	13.5	16**	14.6
Average no. of treatments per day (all days of period reviewed)	3.5	3.1	2.9

* see Table II

** see Figure 1

TABLE II

Distribution of congenital bleeding disorders amongst patients at the Royal Free Hospital (R.F.H.) and the Hospital for Sick Children, Great Ormond Street, (G.O.S.).

Diagnosis	R.F.H.		G.O.S.	
	Registered	Treated Jan-Oct '69	Registered	Treated Jan-Oct '69
Haemophilia	85	53	59	46
Christmas disease	15	12	20	11
von Willebrand's disease	33	9	8	2
Miscellaneous	29	4	2	2
Total	162	78	89	61

Recent Rate of Increase in the Work of the Three Centres

i. Royal Free Hospital

Since starting in 1964, the number of haemophilic patients attending this hospital has steadily increased, viz., 1964 - 5; 1965 - 46; 1966 - 60; 1967 - 99; 1968 - 130; January 1st 1969 to November 4th 1969 - 162.

Where from?

ii. St. Thomas's Hospital

The increase in work load is shown in Figure 2 and below :

	1965	1967	1969 (first 10 months)
1. Average outpatient visits of haemophiliacs per month	10	25	51
2. Average number of inpatient treatments per month (excluding major surgery)			43
			94
3. Total number of haemophiliacs attending at least once for treatment in the year indicated and/or the previous year	49	77	105

iii. Great Ormond Street

The number of patients registered has increased from about 60 to 90 during the last 5 years, despite the referral of patients to adult hospitals (Royal Free Hospital or St. Thomas's Hospital) on attaining the age of about 12 - 13 years. Although figures for the number of treatments in past years are not readily accessible, it can be confidently stated that the availability of cryoprecipitate has led to a very great increase in this respect during the last 2 - 3 years.

Staff Required for a Haemophilia Centre providing approximately 1000 Treatments a year

Director (Consultant)	$\frac{1}{3}$ to $\frac{1}{2}$ time though available full time
Assistant to Director (Senior Registrar or Registrar)	To hold the post for at least one year and preferably two; may have other duties up to half-time but the haemophilia work must be the primary responsibility for which he will be available full-time.
Treatment Officer (e.g. Sister or Staff Nurse)	Full time.
Technician (Technician or Senior Technician I)	2 full time (see note 1, below)
Secretary (Personal Secretary of Higher Clerical Officer)	Full time (see note 2, below)
Teacher	Available as required (see note 3, below)
Medical cover at nights and weekends	(See note 4, below)

Notes

1. Technicians required. We think it desirable that an active Haemophilia Centre serving a part of Greater London should be associated with the corresponding Blood Transfusion Centre in monitoring the potency of the cryoprecipitate supplied by the Blood Transfusion Service in that region. This involves regular assays of the factor-VIII potency of the cryoprecipitate itself at the time of use, and of the response per unit of body weight in patients' plasma after receiving previously determined doses. These assays require the use of plasma from severely affected haemophilic patients which is more readily available in Haemophilia Centres than to Directors of Blood Transfusion

Centres, In many patients, it is essential also to carry out time-consuming tests for the presence of inhibitors of factor VIII or IX. One technician per 1000 treatments per annum should be allowed for the work of monitoring therapeutic materials in the appropriate Haemophilia Centres, in addition to one per 1000 treatments for diagnostic work and the control of therapy.

2. Unit Secretary. The unit secretary is required to act as receptionist and personal assistant to the Director as well as shorthand-typist. Grades below Personal Secretary or Higher Clerical Officer are not suitable.

3. Teacher. In providing hospital teaching for haemophiliacs it is important to remember that these children may have many short absences from school which, in the aggregate, amount to a serious loss of school time (e.g. a third to a half over a boy's life). It must therefore be possible to provide teaching promptly for each school absence of more than a few days. Some haemophiliacs have spent much of their childhood in institutions and are not used to the ordinary school routine; these boys find it particularly difficult to do school work on their own and need much supervision. The difficulties are particularly obvious in adolescence and it is essential to provide a male teacher of strong personality and considerable insight to cope with these boys. We have found that it does not seem to be easy to supply this type of teacher for hospital duties in London from the ordinary resources, and while younger children are usually adequately catered for, we would suggest a special appointment for the older boys in London.

4. 4. Night and weekend cover. Arrangements must be made for selected resident staff to cover these duties backed up by the Haemophilia Centre staff "on call". Allowance must be made for these duties in the planning of junior staffing establishments.

Present Allocation of Staff Time (approximate)

	R.F.H.	S.T.H.	G.O.S.
Director (Consultant)	6/11	4/11	1/11
Assistant to Director	1 registrar (6-month rotation)	Occasional help only	$\frac{1}{3}$ x 2 senior registrars
Nurse	1 staff nurse	$\frac{2}{3}$ x 1 sister	Nil
Technician	1	Nil ($\frac{1}{2}$ seconded from research)	$\frac{1}{2}$
Secretary	Nil ($\frac{1}{2}$ seconded from research)	$\frac{1}{2}$	$\frac{1}{4}$
Medical cover at nights and weekends	$\frac{1}{2}$ x 2 resident pathologists (S.H.O.)	$\frac{1}{2}$ x 1 resident pathologist (S.H.O.) alternating with H.P.	various S.H.O's

? Sister or Staff Nurse for treatments

Additional Staff Required

In order to provide an efficient service of a standard appropriate to a regional haemophilia centre, our immediate needs are for the following additional staff :

	R.F.H.	S.T.H.	G.O.S.
Deputy Director	1 senior registrar* (in place of registrar)	1 senior registrar* or registrar*	1 registrar*
Technicians	1	2	1
Secretary	1**	-	-
Teacher	Special appointment for older boys required, jointly (see note 3, above)		-

* Probably one-year attachments as part of a general training programme in haematology.

** The person appointed would provide help with various social problems (e.g. schooling) arising from other centres.

Therapeutic Materials Needed

An average single adult dose for the treatment of minor bleeds in severe haemophilia is the equivalent of 1 litre of fresh normal plasma - sufficient to raise the patient's factor VIII to about 20%. This activity is contained in approximately 8 packs of cryoprecipitate, but the activity of this material varies widely from pack to pack. Thus for 800 such treatments (doses) per annum, about 6400 packs of cryoprecipitate is required. For surgical and dental cases, higher doses are needed, but cryoprecipitate may be supplemented in these and other cases by the antihaemophilic concentrate prepared by the Lister Institute. Allowing for all contingencies, a minimum of 18,000 packs of cryoprecipitate per annum is required at present for our three centres alone. . .

S.W.T.
Clifton 2,500 p.a. Ayo.
Shenton 1000-2500 p.a. "

20 5
50
1000

For the treatment of Christmas disease, fresh-frozen plasma is required, and this can now be supplemented, when high dosage is essential, by the factor-IX concentrate now being supplied to us in limited quantities by the Oxford Haemophilia Centre.

Dr. K.M. Dormandy,
Pathology Department,
Royal Free Hospital,
Lawn Road,
London, N.W.5.

Professor R.M. Hardisty
Department of Haematology,
The Hospital for Sick Children,
Great Ormond Street,
London, W.C.1.

Dr. G.I.C. Ingram,
Louis Jenner Laboratories,
St. Thomas's Hospital,
London, S.E.1.