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Patron, H.R.H. The Duchess of Kent

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Mr. K. R. Polton, MBE

One of the outstanding occasions in the history of the Haemophilia Society was in 1971 when the Honours List included the name of Ken Polton as a Member of the Order of the British Empire.

This award was an appropriate recognition of the unique contribution he had made to the work of the Society for many years. He was a Founder Member and, at the time of his being included in the Honours List, he had held office continuously for twenty years as either Secretary or Treasurer. For that very long period he had devoted most of his spare time to the Society's work and the welfare of its members and he continued to do so until quite recently.

Some photos from the Society's album of K. R. Polton, M.B.E.

Ken Polton has seen the Society grow over the years to the point where the work has increased so substantially that it can no longer be conducted by officers and other members of the Society working voluntarily in their spare time. Today, it would not be possible for any one person to occupy the key position held by Ken Polton during his time as an officer but the development of the Society's activities and its present effectiveness in serving its members are a tribute to his special contribution.

Before the Annual General Meeting Ken Polton decided that the time had come for him to withdraw from the Executive Committee so we take this opportunity to record our gratitude to him and his wife, Jo, for all they have done for people with haemophilia.

INCORPORATION

We are happy to report that the decision to go ahead with Incorporation received substantial support at the Extraordinary General Meeting held on 23 April. This move will bring the Society into line with the majority of voluntary organisations from an administrative point of view. We would emphasise, however, that it will not affect the work of the Society in any way at all – indeed, you may not even notice the difference. *(Full details about Incorporation were in the Chairman's letter to all members, dated 13 April 1983 – if you did not receive a copy, or have joined the Society since that time, a copy will be sent on from 16 Trinity Street if you ask.)*

CONTENTS

HOME THERAPY – MYTH OR REALITY

Talk given at the Annual General Meeting: 23 April 1983 Professor A.L. Bloom

The title of today's forum is both challenging and provocative. Challenging because it suggests the possibility that home therapy may not be a reality and provocative because on the contrary it suggests that to some people home therapy actually is a myth. Obviously practising clinicians and scientists would prefer to deal with realities and one of my tasks this afternoon is therefore to explain why home therapy appears to be real. At the same time I shall outline some of the logistic and medical problems which may impart a mythical connotation to what is otherwise real. No doubt other myths will emerge during the afternoon's discussions.

When I started to treat patients with haemophilia in the early 1960's the mainstay was fresh frozen plasma. Clearly this was not readily amenable to use for home treatment although programmes which depended on it were initiated. Later when the late Judith Pool developed the cryoprecipitate method for concentrating Factor VIII it was enthusiastically adopted. For the first time treatment of haemophilia A at any rate was readily amenable to straight intravenous injection therapy and on demand treatment at haemophilia centres expanded rapidly. Although storage of cryoprecipitate requires deep freeze facilitie and it is relatively inconvenient to use, it was successfully adopted for home therapy programmes for example by the late Dr. Katherine Dormandy at the Royal Free Hospital and at several other Centres.

Of course the advent of freeze-dried preparations has facilitated the introduction and establishment of home therapy and several reports have outlined the benefits which may accrue (see monograph edited by Jones, 1980). These have been related to reduction of the number of hospital attendances and improvement in education and employment prospects. More difficult to prove has been an actual reduction of joint disability as well as social and psychological benefits from involvement of the whole family in day to day medical care.

Home Therapy in the U.K.

In the United Kingdom the progress of Home Therapy programmes has been monitored by the Haemophilia Centre Directors through its Home Therapy Working Party.

To put the problem in context Table 1 outlines the total number of patients treated in the U.K. during 1981. 2217 patients with haemophilia A received about 28,000 units each of Factor VIII and 368 patients with haemophilia B received about 27,000 units each of Factor IX. Smaller numbers of patients with von Willebrand's disease and carriers of haemophilia A and B were treated. The reality of the situation with regard to home treatment is in fact that just under half the patients with haemophilia A or B are now established on home treatment programmes and the number of patients

Table 1

HAEMOPHILIA
Patients treated in 1981

	Number	Factor u/ patient
Haemophilia A	2217	28,584
Carriers	43	3,186
vWd	282	7,776
Haemophilia B	368	26,769
Carriers	10	4,800

Data collected by UK Haemophilia Centre Directors.

treated has now levelled out. So much so that in fact the Home Treatment Working Party was recently wound up.

What sort of material is now used for home treatment? In Table 2 we see that cryoprecipitate accounts for only a small proportion of material used for treatment and about half of the freeze dried Factor VIII concentrates are used at home for about half of the patients. In fact average use of Factor VIII for patients on home treatment is only modestly greater than that used for those treated at hospital. Thus one of the early fears of over prescribing has not been realised. Of course the corresponding figures for Factor IX are very much lower.

Home treatment for von Willebrand's disease poses a special problem. Cryoprecipitate is usually the treatment of choice in this disorder but dried concentrate may be indicated in some severely affected patients. Although only 9 patients were on home treatment during 1981 they required very large amounts of Factor VIII. Severe von Willebrand's disease can be even more difficult a problem than severe haemophilia.

Thus the reality of the situation is that home treatment exists and is one of the main reasons for the increased use of freeze dried Factor VIII in this country and the declining use of cryoprecipitate over the last few years. From Figure 1 we see that the use of Factor VIII in this country now reaches 60 million units per annum. There is a steady increase in freeze dried concentrates and if we consider that the use per patient per annum in this country at about 28,000 units compares with over 40,000 units per patient as quoted by some authorities for the U.S.A. we see how our projected usage of over 100 million units per annum by 1990 is derived.

Drawbacks

Does the introduction into the circulation of all these blood derivatives have drawbacks? Whilst there is no doubt of the immediate benefit of treatment in terms of daily quality of life and the maintenance of joint function, what about the less beneficial effects?

We are all familiar with the possibility of immediate reactions but these do not pose major problems. Neither do the problems of blood group incompatibility and increased viscosity of blood except in special circumstances. However, the problems of hepatitis, Factor VIII inhibitor and the newly described acquired immune deficiency syndrome pose special risks although not specifically related to home treatment. Thus the percentage of patients who develop antibodies to Factor VIII or IX has remained constant since 1975 at about 6% for Factor VIII and just under 1% for Factor IX. It does not seem therefore that the increased intensity of treatment has increased the inhibitor problem disproportionately. Similarly the number of patients with overt acute hepatitis has remained remarkably constant at about 2 to 3% but there is increasing evidence that more insidious signs of chronic inflammation of the liver are much more common. A survey of this problem is at this moment under way under the auspices of the Hepatitis Working Party of the U.K. Haemophilia Centre Directors.

Acquired Immune Deficiency Syndrome

I cannot end without a comment on one new problem which may turn out to be the greatest myth or the most significant reality of all. I refer to the recently described and publicised acquired immune deficiency syndrome or AIDS.

This condition was first recognised in the United States in 1979-1980 in the male homosexual population of large urban connurbations such as New York, San Francisco and Los Angeles. Its main features (Table 3) consist of a defect in the body's immune mechanisms. Subjects become susceptible to infections. These are not only the usual types such as boils, sore throats, abcesses etc., but characteristically infections by organisms which we all carry but which normally do not cause disease, e.g. fungi and yeasts. In some patients there was defective resistance to a certain tumour called Kaposi's

Table 2

Home Treatment Factor VIII
1981

Material	Total units used Home + Hospital	Units used Home treatment	% used for Home treatment
Cryoprecipitate	6.3m	1.0m	16%
N.H.S.	22.2m	12.6m	57%
Commercial	34.9m	18.3m	52%
Total	63.4m	31.9m	50%
Number of patients	2,217	1,021	46%
Average Factor VIII units/patient	28,584	31,240	–

Data collected by UK Haemophilia Centre Directors

sarcoma and laboratory tests showed various abnormalities and deficiencies of the white cells known as lymphocytes, which normally protect against these inimicable influences.

Table 3

ACQUIRED IMMUNE DEFICIENCY SYNDROME (AIDS)

Defect of immunity to infections
Infections with organisms usually harmless
e.g. fungi
yeasts
Defective resistance to a certain tumour (Kaposi's sarcoma)
Defective laboratory tests of blood white cell function.

Table 4

Acquired Immune Deficiency Syndrome (AIDS)
February 1983 (USA)

Homosexual males in New York,
San Francisco,
Los Angeles
Drug Addicts
(About 1,000 cases February 1983)
Haitian Immigrants in USA
(34 July 1982)
Haemophiliacs
(13 cases February 1983)

At first the syndrome seemed to be confined to homosexual males, many of whom were drug addicts (Table 4) and the cause was not at all apparent. It was thought possibly to be due to an infection e.g. by a virus or to the effect of drugs and for a while amyl-nitrate, which homosexuals use to heighten their sexual awareness, was blamed.

However, two facts developed at variance with the sexual or drug theories. In 1982 the condition was reported in a group of immigrants from Haiti to the U.S.A., few if any of whom appeared to be either homosexuals or drug addicts. Then, in 1982 also, the condition was reported in a few haemophilic patients in the U.S.A. By February 1983 thirteen cases have been reported amongst American haemophiliacs only one of whom was an admitted homosexual. Eight of these patients have since died. I am unaware of any definite cases in British haemophiliacs although cases are occurring in British homosexuals and it is rumoured that one of these has haemophilia. The Haemophilia Centre Directors' organisation is conducting a comprehensive survey of the prevalance of AIDS-related abnormalities in our haemophiliacs population so that firm data may be available in due course.

In addition physicians in the U.S.A. and Great Britain have now turned their attention to the 'healthy' haemophiliac population. Evidence of impaired lymphocyte immunity has been detected in 'ordinary' haemophilic patients but this occurs also in many other conditions. Nevertheless it is probable that like most newly described disorders less severe or early forms will be detected and many research projects are under way in attempts to elucidate the extent of these changes.

What is the cause of the disorder? This is still quite unknown. It could simply be a reaction on the part of our immunity mechanisms to the long term infusions of other people's plasma proteins. Perhaps there is some unknown constituent of blood products which is responsible, but the occurrence of the condition in homosexuals, drug addicts, immigrants from tropical climes and in recipients of blood products makes the transmission of an infective agent the most likely cause.

How should we react to this development? It is important to look at this in perspective. All the commercial Factor VIII producers are taking steps to select their blood donor population more rigorously, but it is worth reflecting that thirteen cases amongst 20,000 haemophiliacs in the U.S.A. means only one expected case amongst 2,000 haemophiliacs in the U.K. Thus American concentrates have been used in this country for many years and the AID syndrome is not overtly prevalent here. Indeed evidence from the incidence of hepatitis does not lead one to believe that concentrates prepared from British blood are necessarily safer, in this latter respect at least. Although it is prudent to keep an open mind, the use of factor concentrates has revolutionised the lives of many sufferers from haemophilia A and B and it does not seem reasonable to curtail treatment at the present time.

Supply of Blood Products

The supply of blood products is not the only important aspect of haemophilia management and other facets of care are important. Nevertheless the supply of Factor VIII concentrates has figured prominently in our deliberations. At the end of 1982 the Central Blood Laboratories Authority was established to assume the management of the Blood Products Laboratory at Elstree and the Plasma Fractionation Laboratory and the Blood Group Reference Laboratory in Oxford. It is planned to expand production of Factor VIII and other blood products to meet all our projected domestic requirements so that this country will become self sufficient in blood and blood products.

Thus in the end of course the genes controlling the production of Factor VIII and Factor IX will be isolated and introduced into fermentation organisms thus relieving us of dependence on donated blood. But let us not be too complacent. As with hepatitis and AIDS, so the new technology will bring new and unexpected complications. Hopefully our younger colleagues will rise to the occasion and find new solutions to these and to older problems.

Professor A.L. Bloom

Reference

P. Jones (Editor) Haemophilia Home Therapy. Pitman Medical, London, 1980.

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ANNUAL GENERAL MEETING – 1983

Question and answer time

Panel: Rev. Alan Tanner (Chairman)
Dr. Forbes
Professor Bloom
Sister Fountain

Rev. Tanner: By popular request we shall be dealing with a subject of current interest – home therapy!

Question 1. I believe that home treatment would result in less usage of Factor VIII because early treatment enables lower doses to be used. However, I understand that patients going on home treatment are tending to use more Factor VIII.

Prof. Bloom: I am not at all surprised. I think there are 2 reasons why. When people have treatment at home it is more readily available and they are more likely to take advantage. When one had to come up to hospital one was inhibited from coming up but now at home all you have to do is get it out of the fridge and give it. In the early and mid 1970s supplies of Factor VIII concentrate were increasing and they are still increasing. One of the reasons is that it is effective and supplies are more readily available and people have used rather larger doses because they have found larger doses are more effective.

Sister Fountain: One of the reasons is because of prophylaxis. When at home one may have a dose of Factor VIII when one normally would not be bothered with one if one had to come up to hospital. Also peopl are put on long-term prophylaxis. This would not be practical if one had to come into hospital. When children were put on prophylaxis treatment before home treatment was introduced it meant that children needed to be brought into hospital 2-3 times a week.

Dr. Forbes: When home treatment started the average usage was about 40,000 units per year. It is now much higher. We are now using 20-55,000 units per patient per year. Implications of cost are quite great. We still do not know how much to give for one individual bleed. Usually we give 1-2 ampoules. Some groups are looking at Factor VIII per vial. When we have a lot

of Factor VIII the usage will probably increase.

Question 2. What effect does home treatment have on individuals? Does home therapy make an individual independent or does it merely move his problem from the centre back to himself?

Sister Fountain: A lot of people have this in mind. Home treatment changes one's life style quite a bit. Usually patients are now independent and have a feeling of controlling their own situation and if they have a bleed they know they have got something nearby to treat it with but at the same time they must be able to have somebody to help them if they have a very bad bleed. A close liaison must be kept with the Centre; certainly where parents are concerned with young children. If they are isolated they must be able to telephone at any time to ask small questions which they may not want to ask their own doctor as they feel he may not have time for such questions. As long as they have a fairly close connection with the Centre then they do not feel isolated.

Dr. Forbes: It depends on what you mean by independent. A lot of people on home treatment say they are able to become more independent. They can go to school, keep a shop, etc. The main point of view is that it has a very dramatic effect on one's life. Undoubtedly, it does shift the basis of many day to day problems back to the patient but it is still important to keep in touch with the Centre so a telephone is essential if people are on home therapy.

Prof. Bloom: I am sure that independence has increased. I am not too sure if it is at the expense of the Centre. Patients would be well advised to keep in touch with the Centre. Home treatment means increased responsibility for the doctor. Increased independence means increased activity which means increased usage.

Sister Fountain: When I first came to St. Thomas' I found that patients went on home treatment and it took some time before I knew who people were and got to know their special problems. I do know other nurses who have had the same problems.

Dr. Forbes: Many patients used to be very dependent on the Centre. We still have patients who come up every day. To change this would now be impossible.

Member of audience: I have been on home treatment for nearly 10 years. I still get cheesed off at having to be patient and doctor and having to decide whether the bleed is bad enough for that amount of Factor IX and sometimes it is difficult to get advice from the Centre because the staff change all the time. They say you are in charge if you are on home treatment but I still want the Centre to take responsibility. I just want to be the patient.

Sister Fountain: It's a two way thing. You know you can always ring.

Member of audience: Normally it is easier when you are talking to a patient across a desk because you can examine them. When a patient 'phones up it is difficult to assess the situation and even more difficult because you know they will rely on your advice even though it may be wrong.

Question 3. What are the panel's views on the patient's sense of failure if he fails to treat himself successfully and in consequence has to attend the Centre?

Dr. Forbes: Failure is not an important point.

Prof. Bloom: I agree this is not an important problem. It depends on the individual; some patients are more competent than others. Doctors and nurses do not regard them as failures.

Sister Fountain: It is very important that they do not feel like this. They can ring up at any time.

Dr. Forbes: If a patient 'phones up and they have a headache I have a legal responsibility to them. They must come in to

see me. Often they 'phone the Sister. The doctor has to see the patient, even though the problem is often trivial.

Sister Fountain: Merely the fact that somebody has 'phoned means they are worried and need to be seen.

Prof. Bloom: If somebody is worried they should 'phone and they should probably come up. One has to play safe.

Dr. Forbes: How many actually 'phone up their own family doctor?

(Nil response from audience)

Sister Fountain: A lot of people live a long way from Centres. Some of our patients live many miles away. We have to make sure that we get back up from local hospitals near the patient. Another important point is the question of the staff moving and patients never getting the same doctor. Usually the Sister is the most suitable person to talk to but there is the problem of maintaining 24 hour cover.

Prof. Bloom: This is no different from any group of people who are ill. There are 2,000 patients in this country with severe haemophilia. There is always some suitable person, i.e. Haemophilia Centre nurse or director. Junior staff are always rotating because they have to because of the system of training we have in this country. We can never implement permanent junior staff.

Member of audience: The main problem is of 24 hour staffing and how to get trained staff at night if we have a problem. The house physicians' knowledge is now limited as there is so much home treatment. The house physician who is in charge at nights does not know much about haemophiliacs.

Sister Fountain: At night and at weekends, when most people find problems, the house physician or senior house officer has contact with the Centre, i.e. they can 'phone the consultants for advice.

Rev. Tanner: Many people have told me that they are beginning to get the feeling that they are being pushed into home treatment. If they are not on home treatment they feel they have failed in some way.

Sister Fountain: I understand that side of it very much. What is perhaps easy for some is not easy for others. The actual putting of a needle in is very difficult for some patients. That is the bit most people are worried about. If they can get over that feeling then most people overcome any other problems. The problems come with parents of young children. It is hoped that parents of young children will be trying them on home therapy. This assumption is made from the earliest days and some parents are rather worried by that. They are worried by how their children react to experts. They are worried that within the next two years they may be expected to treat their child at home, they have serious doubts as to whether they are going to be able to do it.

Dr. Forbes: Parents should not feel that they are going to be pressurised. If you are willing and able to cope then that is fine, but hospitals should not pressurise you. If you feel you cannot do it, then do not be pressurised.

Member of audience: I have a 6 year old haemophiliac. Fortunately we have a very good Haemophilia Sister and have a good routine at hospital and it was fairly easy to go on home treatment. However, there is always that thought there of "what happens if I fail to get the needle in and do not get my son to hospital on time?" The pressure on the whole family is considerable.

Sister Fountain: With young children it is difficult because of the veins. With all the best will in the world it is the veins that are the problem. The experts have difficulty so parents should not be surprised if they have difficulty.

Member of audience: We have been discussing home treatment by the adult haemophiliac himself, or by the parent of the haemophiliac, in terms of concentrate. Could I ask the panel if they could explain treatment to cover the management of analgesics at home. I would like to hear what they have to say about analgesics.

Dr. Forbes: One of the unsolved problems in haemophilia. We still see up to 10 per

cent of patients in very big Centres with analgesic problems, i.e. abuse of some shape or form. They have various reasons for abuse, often give morphine etc. We see a problem with DF118. This is a problem we cannot resolve at the moment. If an analgesic problem needs to be faced, it is not terribly helpful to wean them off analgesics. If someone is in pain we have to treat them for pain and they have to live with the long term consequences of that.

Prof. Bloom: The problem of analgesia abuse seems to be greater in the cities. I have not got any patients who are dependent on analgesics although a few of them need them. I am sure none of my patients are not dependent.

Sister Fountain: In our Centre we have 2 people who have a serious drug problem. They are controlled on a certain amount of drugs. All their requirements have to go through the department. The general problem is what can I take for pain? The most sensible way is to treat the bleed before the pain. If pain, in particular pain in the joint, occurs they should treat the pain and find the cause. In the last 2-3 years I have seen that particular problem going on.

Member of audience: On Monday the BBC Horizon programme has a programme on AIDS -- called the Gay, Black, Haemophiliac's disease. This unfortunate association in the Radio Times must be upsetting for haemophiliacs. What is the situation and what are the reasons behind this?

Prof. Bloom: It is unfortunate that haemophilia has been linked with AIDS. Apart from that we must not overlook the AIDS problems. One of my patients may have a mild form of it. Some patients show laboratory changes. Laboratory changes do not mean that it is a serious disease. I do not know of any haemophiliac with AIDS in the U.K., France, or Germany. I do not think we need to get over-concerned about this. At the present time it would be absolutely wrong to curtail treatment.

Dr. Forbes: I would like to underline this. The problems in the United States are not prevalent anywhere else. It has not occurred in Britain or in Western Europe. Why it should happen in this way in the United States is very strange. The patients who have succumbed to it have had infections of a very unusual type. We have seen this kind of thing before in people. It is possible that some new disease has been imported. Why is it not present in Europe? How many of you have heard of this disease? Is it something that has been worrying you?

Member of audience: We were not warned about AIDS but we are concerned about public reaction to this programme. I feel it will get too much public attention. The same thing happened with hepatitis B when it started and it got front-page cover 10 years ago in the Daily Express. I do not know what one can do to change the course of journalism.

Prof. Bloom: The concern has been that transmission has been from blood or blood products. The relevance is whether the transmission of this disease is by blood and associated blood products. This is what it is concerned with. If it is a transfusible agent it is strange that it has not happened in Western Europe. They are only assuming that it is transfusible by blood. Not everybody who gets blood will get the disease.

Member of audience: The first time I heard about AIDS was in connection with haemophilia on a programme to do with sexually transmitted diseases. It was the first time I heard it could be linked with haemophilia. The programme was not very informative.

Rev. Tanner: Can the panel advise on the disposal of used equipment in home therapy? What about concentrate bottles?

Sister Fountain: The best way is to take used things back to the Centre and they will get rid of them properly for you. I certainly do not advise patients to dispose of their own things. You must not put

bottles in dustbins; if you do you will get into a lot of trouble from the local council. You should definitely take things to your local Centre or local hospital. Arrangements can usually be made. Needles should be put into a box and when full taken to the Centre.

Dr. Forbes: We give patients a "sin bin". There is a very great hepatitis risk from dirty needles to the family if not disposed of properly.

Prof. Bloom: There is no such thing as a hepatitis free concentrate. The concentrates must be handled with care.

Sister Fountain: This problem also concerns staff. I wanted to know what the risk was. I have now been convinced that the risk is very slight. The best way is to be properly trained and to be very careful with all equipment. One should be doubly sure not to leave concentrates lying around.

Member of audience: Is there any risk from open wounds on the families?

Prof. Bloom: Yes, there is a risk. The evidence shows that when people have been exposed there is a high risk that they may develop non A or non B hepatitis. One individual pack of cryoprecipitate is a risk even though small. About 1 per cent of the population carries hepatitis B. They may not have been aware of carrying the infection.

Question. Could a member of the panel tell us about the availability of home treatment for people with inhibitors.

Prof. Bloom: I do not put patients with inhibitors on home treatment. In individual cases there may be a case for home treatment. As far as I am concerned none of my patients with inhibitors are on home treatment.

Dr. Forbes: We do not treat inhibitor patients with home therapy.

Sister Fountain: It is dependent on the level of antibody and how the antibody reacts to antibody and in a few cases we do treat patients at home. Ordinary Factor IX concentrate for small bleeds works fairly well. However, home treatment does not always work.

Member of audience: Could the panel comment on the areas where home therapy would be particularly well or less well indicated?

Dr. Forbes: All facilities are better in major Centres. We in Glasgow cover the Western Isles of Scotland. We have one or two patients on home therapy and we send material out by post to them. We have trained and instructed the local General Practitioner. The local hospital also knows what to do in an emergency.

Prof. Bloom: We too have patients who live in remote areas and we have a few patients who are successfully on home treatment. They get their material either by a special delivery to a nearer hospital or by a blood transfusion van. They seem to cope. It is impossible to deal with remote areas.

Sister Fountain: No reason why not. It is quite important to have a first aid measure for a major bleed. Some arrangements can be made with a particular General Practitioner or a local hospital. It should not be too difficult. Posting of supplies has to be done in some cases. However postage sometimes gets held up but this can be got around. Patients who live in remote areas do not have to be left out. Sometimes the domicillary nurse has taken part in home treatment programmes. People who live 60 miles away from London have a visit from the nurse and this works very well.

Question. Could Professor Bloom tell us about the availability of home treatment, since it appears that those who could benefit the most -- i.e. in remote areas -- are unlikely to be recipients?

Prof. Bloom: Very difficult question to answer. I have not seen any statistical data on this. Peter Jones and other members of the Home Treatment Working Party thought that there was an increase in people being put on home treatment and that the Home Treatment Working Party had done as much as they can. Do you think we should re-open the Home Treatment Working Party?

L to R, Prof. Bloom, Dr. Forbes, Sister Fountain and Rev. Tanner.

From Floor: About 45 per cent of patients are on home treatment. This is still leaving just less than 60 per cent. It does not seem to be very many.

Sister Fountain: What percentage of that 60 per cent is young children? We have several under the age of two. Every time a new patient is found that is another patient not on home treatment.

Prof. Bloom: We shall never get 100% on home treatment.

From Floor: Are the Working Party satisfied with their review clinics in connection with home therapy programmes? Whilst home therapy is readily available to almost anybody who asks for it, in my view, the review clinics to ensure that treatment is effective are not always taking place.

Dr. Forbes: This is difficult to answer. How does one maintain standards? We run review clinics. Part of the conditions of Home Therapy are that they come for review. If they do not come they are taken off home therapy.

Prof. Bloom: I would like to be able to identify where the local haemophilia Centre Director does not carry out regular review clinics. I see my haemophiliacs every six months at least — even if they have no complaints. If you can identify an area where this does not take place we cannot do anything about it but the Society can.

From Floor: How much training is given to those not on therapy with regard to allergic reactions?

Sister Fountain: My policy is always to teach about reactions to treatment. There are usually fairly mild reactions to concentrates. Of course people on home care must understand what reactions are and know how to deal with them.

From Physiotherapist in audience: I wonder, as a Physiotherapist treating patients on Home Treatment, whether they are reviewed enough. We are getting young children who have a severe joint problem at the age of 9 or 10. When Home Therapy becomes more prominent I hope not to see children immobilised at a young age.

This is a very difficult question to answer. No doubt, a certain number of children and adults are still going to get chronic orthopaedic problems in spite of apparently adequate home therapy. Even in the best regulated circles. You need to get a balance of the amount of Factor VIII given against the side effects against the risk of developing an orthopaedic problem.

I would just like to comment about our Orthopaedic Care. At our review clinics the Director of the Centre, the Physiotherapist, and the Orthopaedic Consultant all see the patient every 6 months or at least once a year. This is a combined effort. The patient has quite some time to go into any problems which they are having.

Combined clinics seem to benefit all much more. They get problems sorted out.

NEW APPEAL RIGHT ON MOBILITY ALLOWANCE

DHSS Press Release August 1983

Some disabled people will gain a better chance to turn their mobility allowance into a car on the road, as a result of a change announced today.

The Mobility Allowance regulations are being amended by a new provision*, operative from 29 August, which will extend the right of appeal for people who claim Mobility Allowance. In addition to the right of appeal against a decision not to award the benefit, recipients will be able to appeal against the period of award if it is for less than the maximum period, i.e. until the recipient is 75 years old.

This will be of particular help to those people who would like to lease or buy a car through the Motability scheme. An award of 4½ years is usually required before a person can buy a car through the scheme and an award of 3-4 years is needed for a leasing agreement (depending on the type of car). Thus if someone is awarded the benefit for periods shorter than these, he or she is effectively excluded from the Motability scheme.

"I am very glad that we have been able to introduce this change," commented Tony Newton, the Minister for the Disabled today. "It should enable more people to take advantage of the Motability scheme, thus increasing their mobility which is so important in helping disabled people to lead an independent life."

Note: Mobility Allowance is a benefit which can be awarded to people aged between 5 and 64 years who are unable or virtually unable to walk and once the allowance is in payment it may continue until the person is aged 75. The weekly rate is £18.30 (£19.00 from November 1983).

Motability is a scheme under which recipients of Mobility Allowance can have the benefit paid directly to Motability and lease or buy a car from them in return.

*The Mobility Allowance Amendment Regulations 1983. SI 1983 No. 1186.

MOBILITY ALLOWANCE/ ATTENDANCE ALLOWANCE

Increasing number of people with haemophilia are now winning the long battle to obtain their benefits. The Co-ordinator is most willing to help with the preparation of your case, and representation if that is possible. Your attention is also drawn to manufacturers' discount schemes available to people in receipt of Mobility Allowance — as well as the **Motability** scheme. Motability offices are at:

Boundary House
91-93 Charterhouse Street
LONDON EC1M 6BT.

BADGES

A reminder that SOCIETY BADGES are available from 16 Trinity Street. They cost 50p each, plus 12½p postage for up to 6 badges: thereafter add a further 12½p to cover additional postage.

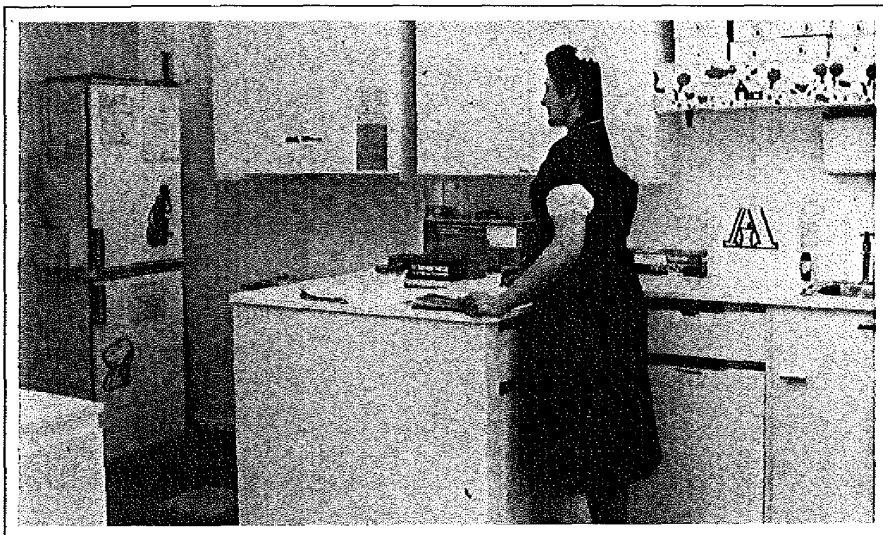
(We will soon have Society 'T'-shirts as well — so you will be well equipped for the next Marathon, or Diminishing Tea Party!!)

The Haemophilia Society

REMEMBER you can "Telephone us anytime"

We are now on a telephone answering machine which is available whenever there is no one in the office to receive your call personally. **Note that the office is now staffed Monday to Friday from 9 am to 5 pm.**

'No point in ringing — there is never anyone there!' — NOT ANY MORE!!



Sister Marion Gregory at work in the new haemophilia unit at The Children's Hospital, Birmingham.

CHANGED (or changing) YOUR ADDRESS?

Please let us know of your address change as soon as possible. We lose contact with so many members each year when we are not told of address changes!!

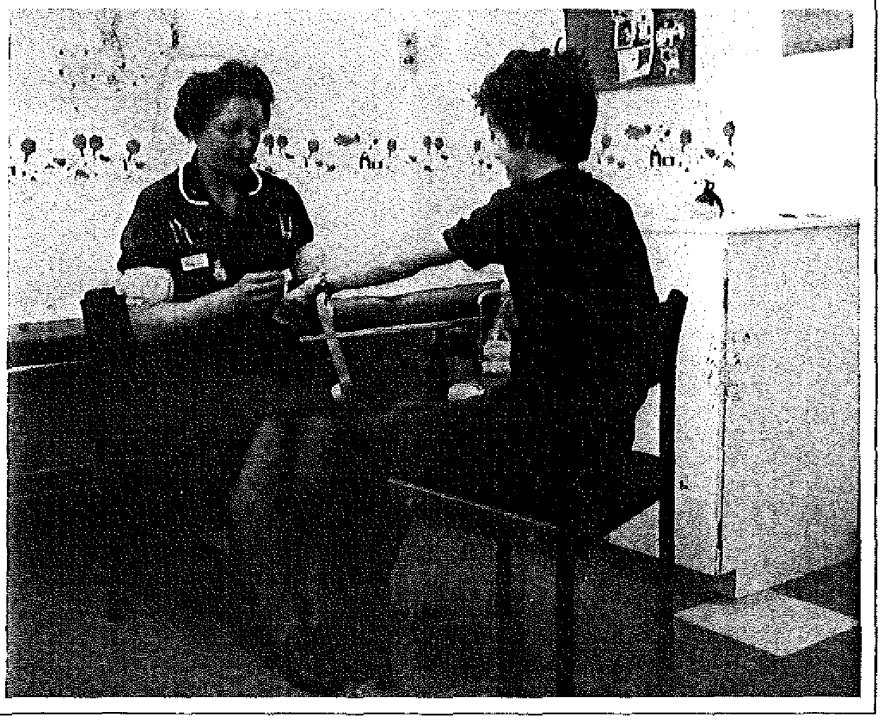
★★★★★★★★★★★★★★★★★★★★★

INSURANCE

Remember that the Society can help you to obtain cover for life, holiday, car and other forms of insurance at highly competitive rates — and the Society earns the commission!

THIRTY YEARS OF HAEMOPHILIA or A TALE OF BUT'S

Elizabeth E. Mayne, Consultant Haematologist, Royal Victoria Hospital, Belfast



My name is William James Cornelius Anthony Pat!
My Dad says he's to blame for most if not for all of that.
I was born on a sunny day in the merry month of May
In the year one thousand, nine hundred and fifty three.
Sure, all thought I was perfect, as far as the eye could see.

The weeks and months flew past BUT bruises came thick and fast,
The Doctors came and said, all is not just as it seems,
We think your Mum has passed to you a defect from her genes.
Your clotting is not great, in fact you're missing Factor Eight.

★★★

The next few years I didn't really mind,
The doctors and the nurses were most awfully kind,
BUT, up and up I grew — No — still nothing new
Just the plasma and the pills and the endless rest in bed.
The POP casts, sometimes they seemed as heavy as lead.

The Doctors, always changing, but going to the greatest pains
To pump their little potions into my poor old veins,
Then, one day I was told I could have something new and cool,
Thanks to a Yankee lady, whose name was Judith Poole.

I got in such a state — at last — the missing Factor Eight,
The pain just went — thank God for cryoprecipitate!
My life it was transformed — imagine the relief
No more ghastly bleeding when the dentist pulled my teeth.
My joints were quickly treated, the pain soon went away,
I was off to school all within the very next day!

★★★

By 1973 I'd reached the age of twenty
I was doing a little work but mostly my life was joy and full of plenty.
BUT, an episode occurred, I'm unhappy to relate,
The cryo didn't work, I got in such a state!

What had happened to the stuff, perhaps it wasn't strong enough?
My blood was checked and Oh, Unhappy Fate,
I was told I had developed an anti Factor Eight.
I asked them, please tell me why; all I got was
"your response is very high"
No treatment for a while, back to rest in bed.
I used to wake up in the morning and I wished I might be dead.

★★★

BUT then, despite no treatment, I had two years of peace,
To my very own surprise, my bleedings did not increase.
Then I had a tragedy, I fell down on the farm,
I broke my leg in two and got a haematome the full length of my arm.

I said I'm in an awful stew, please tell me what you're going to do,
The Doctor said, "Don't worry Pat, we'll soon take care of that,
We've got some new 'eight deficient' super glue!"
It surely did the trick and I wasn't even sick.
I asked them to explain how something without eight, could work inside **me** just the same.
I was told, it's full of Factor Nine which bypasses the need for eight, along the clotting line.
It was called FEIBA, and sure, I felt things were now going my way.

BUT, within a month or so I turned an
awful shade of yellow,
I didn't feel quite so smart a young fellow.
As usual, the doctors checked my blood
and then they said to me,
You've got the hepatitis, the type not A
nor B.

★★★

Thankfully, no more special treatment
did I need,
Until one year later I got another great
big bleed,
This time I was treated with something
from a pig!

Now I'm not a fussy type but I'll say to
all of you,
I wish those Doctors knew just what
they're trying to do!
BUT I'm happy with my 'pig' when my
bleeds are whopping big.

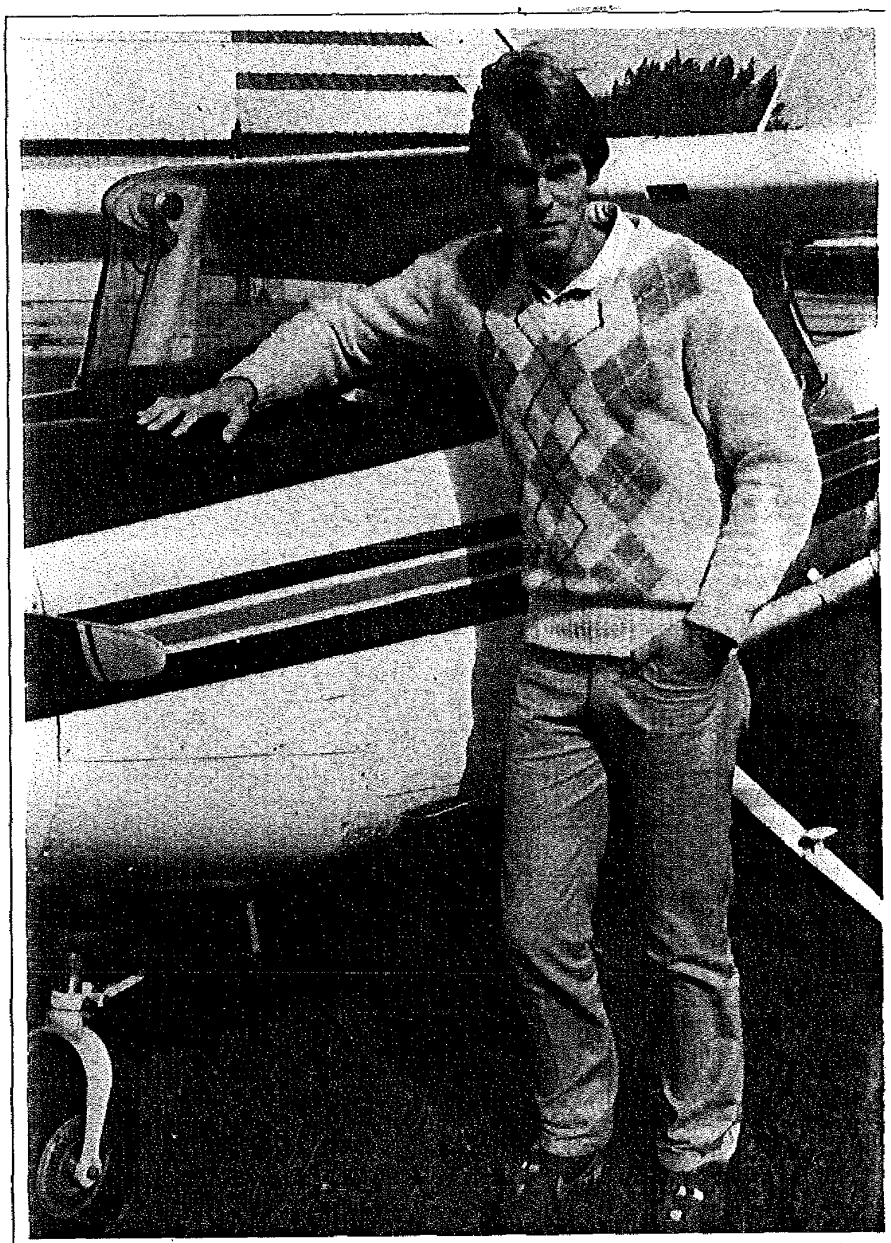
Last year I joined a scheme to help the
local team,
It was called a double blind cross over
trial,
Its aim was with **another** potion to keep
my joints in better motion
And to improve my rather cramped life
style.

I was summoned to attend just as soon as
one got big and didn't bend,
I waited and I waited but my joints just
didn't bleed.
They really are so fickle, just when my
Doctor was in need.

At last I got one in my elbow and I had
my trial run,
I got another in my knee and my Doctor
had more fun,
There was no great dramatic good BUT
maybe I wasn't in the mood,
Perhaps I'd had enough of all this funny
stuff.

I bear the Doctors no ill-will, rather the
reverse,
Without their help I'd be more than 100
times worse,
BUT I wish with all their brains and
skills
They could find a cure,
So I wouldn't have to endure —
It would give me such a thrill if the cure
could be a **little tiny PILL!**

★★★★★★★★★★★★★★★★★★★★★★



Bill Somers (aged 19), who recently won a Douglas Bader Flying Scholarship. Bill's father is a Squadron Leader, his sister Chairman of our Oxford Group, and his brother a BA pilot! Bill, a moderate haemophiliac, is studying at Swindon College and we all say 'Well done' to him!

A MEMBER ACHIEVES HIS 90th BIRTHDAY

Yes, it is a wonderful thing to live to this ripe old age, and equally because of his medical record.

Frank Wycherley of Gatley had the usual trials with joints, cuts and gashes severe and less so, in a South Cheshire country village. His joint troubles were attributed to a rheumatic condition and copious hot fomentations were applied with damaging results.

F.W. played football once, for his county school, and beforehand, he fell on his bare knee, got blood poisoning; a leg amputation was talked about and it was a six months' job, finishing his school education at the age of sixteen.

Mr. F. Wycherley

F.W. then served an apprenticeship in the light electrical engineering trade, and when war broke out, he followed his two brothers and joined the army at the age of twenty-one. He lasted 110 days in the Cheshire Yeomanry, and was discharged after a tooth extraction, despite lying low in the billet for several days to escape the unwanted verdict.

Fit again in a month, F.W. went to Liverpool and joined the mounted section of R.E. Signals and served two periods in France, and finished on the Somme with violent toothache. Extraction of a tooth in the Medical Officer's tent followed, and he had to have arm-to-arm blood transfusion in a surgical ward in Rouen, and to Blighty. Later he was discharged after a finger gash.

It was late in 1916 when F.W. went to Manchester and did forty-one years in the electrical manufacturing industry, escaping in the twenties a staff blitz by being on the 'King's Roll' for discharged soldiers.

During 1936 F.W. needed dental treatment badly and heard of the Sheffield 'Egg White' Timperley Treatment for Haemophiliacs. He went to Sheffield for a month, got rid of the five teeth and continued with the beneficial treatment until the serum production ceased during the war.

In February 1939, F.W. joined A.R.P. (Air Raid Precautions), as a warden, but came to grief in training with a leg injury, damaging the crural nerve and after severe pain for some weeks, was discharged as unfit, before the second war started.

Since retirement at 65 in 1958, F.W. survived two severe internal haemorrhages, one from bleeding ulcers, necessitating emergency treatment, and his first surgical operation, which was successful though touch and go.

Since 1951 F.W. has been an outpatient at the Manchester Royal Infirmary, where 'Cryo' is his lifeline.

Now at 90, F.W. hobbles around his small garden, and has help in growing flowers and vegetables. He has given up his hobby of coarse fishing, but still does nut and bolt repairs in his workshop.

F.W. is still a car driver on local journeys and says he owes his long life to physical moderation, to the hospitals and last, but far from least, to his devoted and active wife, who is an octogenarian.

YOUR CENTRE

THE LINCOLN HAEMOPHILIA CENTRE

Before 1981 a huge area of eastern England, comprising the counties of Lincolnshire and South Humberside, had no Haemophilia Centre of its own. Patients frequently travelled over 60 miles to Sheffield, Nottingham or Derby for treatment. It was not unknown for local hospitals to commit grave errors in managing haemophilic bleeding due to unfamiliarity with the disease and ignorance of the service provided by Haematologists in distant hospitals.

Since 1981 the situation has improved greatly with the opening of Lincolnshire's own Haemophilia Centre, based at The County Hospital, Lincoln. The Centre serves over 60 haemophiliacs, about half in the 'severely' and 'moderately severely' affected category. Housed in Victorian buildings the Centre has the great advantage of being just inside the front door of the hospital. Patients can be driven to within a few feet of the treatment area and porters are on hand to assist with chairs and advise on car parking etc. A major disadvantage is that we share a treatment area with patients attending for blood sampling and treatment of other blood diseases.

The Centre provides a round-the-clock service with two Consultant Haematologists available to supervise treatment and give advice to outlying hospitals. Sister Brenda Brown, our Haemophilia Nurse, is responsible for administering therapeutic material, training patients for home treatment, issuing supplies of anti-haemophilic factor and home treatment packs. She has also become an expert Agony-Aunt specialising in social problems of a delicate nature.

The interests of all members of the team overlap in particular with our Social Worker, Mary Dundon. Mary has become a member of the British Association of Social Workers Special Interest Group (on Haemophilia) and has represented Lincoln at their conference in London. She has become particularly expert in educational matters and arranging financial assistance for haemophiliacs who have become unemployed as a result of their condition.

By no means the least of our blessings is an active local branch of the Haemophilia Society with Mr. Leslie Mumby as chairman and Mrs. Sue Cooke, secretary. The group is usually well attended, meeting on the last Sunday of every month in the Nurses' Home. Recently it has concentrated on three main areas: providing a forum to discuss developments in the treatment of Haemophilia; fund raising for the National Society and encouraging and assisting needy families in our own area. Recently it gave financial help towards a holiday on the coast and also purchased vital equipment for the laboratory. This year six year old Ben Harrison won the Brendan Foster award for his swimming as a result of encouragement from the local group.

As a new Centre we are trying to incorporate all the best features of larger, well established Centres. This we will continue to do after we move to new premises in the District General Hospital, due to open in 1985, with the aid of our enthusiastic staff, loyal patients and the generosity and support of our local branch of The Haemophilia Society.

D. R. Prangnell & M. I. Adelman

Dr. Prangell, Director of Lincoln Haemophilia Centre with his staff

WORLDWIDE

WORLD FEDERATION OF HEMOPHILIA CONGRESS – STOCKHOLM 1983

Andy Cowe, Chairman Scottish Group

I was delighted to be chosen, along with Diane Lewis and Bob Emery, as a representative of the Groups to attend the WFH Congress in Sweden. Until June of this year, I had no personal experience of WFH meetings, and it was with a sense of stepping into the unknown that I set off for Stockholm on 26th June 1983.

What I found at the Congress can best be described as a week of total immersion in haemophilia. From the journey there until the last farewell at Heathrow 6 days later, the subject of every lecture, the illustration of every exhibition and the topic of almost every conversation was haemophilia or a related theme. During the Congress I learned a lot, I became even more aware of my lack of understanding of some topics, and I took away a lot of new ideas and thoughts, some of which I hope I can share in this article.

The first problem of the Congress was planning one's programme for the week. The splendid Karolinska Institute has three lecture halls, each of which was in use for the whole week, so each lecture had to be chosen from a choice of three. In addition, there were the famous "poster sessions" (of which more later) which had to be viewed. Another competing attraction came from the display stands of the various pharmaceutical firms, so every minute in the Institute was taken up in some way or another.

The Congress was officially opened on the Sunday evening in the beautiful Berwald Hall where we were entertained with music and folk dancing. We then moved to the Biologiska Museet, a largely open-air biology museum where we were given a meal and an opportunity to meet other delegates.

The Congress proper began on Monday morning. I attended a symposium on the Psychosocial Aspects of Haemophilia, which comprised five papers dealing with the common theme of understanding how the individual and the family come to terms with haemophilia. The whole Congress then assembled for two plenary lectures. The first was on Bioengineering of Blood Derivatives, and proved far too technical for a mere layman like myself; but the second on the State of the Art – Von Willebrand's Disease, increased my knowledge of this condition considerably. Each delegate was given a tape recording of this lecture courtesy of Cutter Laboratories Ltd., and for those interested, it is well worth hearing.

On Monday afternoon, the symposium I attended dealt with Health Delivery Care. For me this was perhaps the most interesting session of the Congress, dealing as it did with a wide variety of topics at a level which was more suitable for the layman. Of the ten presentations, I particularly enjoyed the three mentioned below. Newcastle's own Maureen Fearns presented the Illustrated Haemophilia Guide, which has been prepared by WFH's Task Force I Nursing Subcommittee. This is an excellent teaching aid and with its visual format overcomes language barriers, giving the book worldwide application. Maureen achieved further fame as her appearance at the lectern was used by Swedish Television in a lengthy news item on the Congress that evening!

Dr. P. H. Levine read a paper proposing that WFH undertakes a classification of comprehensive care centres all over the world to provide a standardised frame of reference for those using and providing haemophilia Treatment Centres. This is an interesting idea, but the introduction and maintenance of such a register would present enormous difficulties.

Y. Laurian of France presented a lively video film illustrating the benefits of carefully supervised and managed physiotherapy in improving joint mobility in a young haemophiliac.

EXCHANGE HOLIDAYS 1984

After a lapse of a few years, the Society will once again be assisting young people to obtain exchange holidays with families all over Europe. There is already the possibility of an exchange with an 18 year old in Florence and the Irish Society are keen to have exchange holidays with the United Kingdom. **Please write to the Co-ordinator if you are interested in this scheme.**

The session concluded with reports and views on the role of the paediatrician in haemophilia patient care. Dr. Peter Jones (Newcastle) made a strong plea for the inclusion of paediatricians in haemophilia care teams, particularly as they had not been explicitly mentioned in Dr. Levine's proposed classification.

On Tuesday morning the first session I attended dealt with the availability of antihaemophilic factor in its various forms at the worldwide level and also in certain selected countries. A session on Psychomedical Views was next on my agenda. Three interesting papers on patient education, group therapy and genetic counselling stimulated a lively discussion.

The afternoon symposium started with a description of a particularly South American aspect of haemophilia care – the haemophilia home, or Casa de Hemofilico. These homes are a sort of residential comprehensive care centre, where haemophiliacs from the remoter parts of countries like Brazil and Argentina can stay while they are assessed, given therapy or rehabilitated.

From the situation in these less privileged nations our attention was turned to the Role of the Professional Executive Director of Haemophilia Societies. The platform was taken by the only three professional Haemophilia Society administrators known to the World Federation, one of whom is, of course, David Watters from our own Society, the others being from the USA and Canada. The three speakers ably demonstrated the benefits which accrue to Societies through professional administration and pledged their support to each other and the aims of WFH.

On Tuesday evening, one of the social events of the Congress took place in the form of a boat trip to the island of Waxholm. Here we visited a 16th century fortress which is now used as a military museum. A traditional meal was served, then folk dancers demonstrated their art before encouraging their guests to join in with some fairly boisterous, not to say undignified dancing, which was greatly enjoyed by all. This participative atmosphere encouraged a number of delegates to continue the party on the boat trip home, with singing and dancing en route. I was lucky enough to be treated to some flamenco from the Spanish delegation, while on the other boat I believe that a Scottish Cockney entertainer co-ordinated the singing!

Wednesday saw the General Assembly of WFH – the business meeting of the Federation. Three new member countries were welcomed into the Federation. Reports were received from all the major Committees of WFH, and thanks were expressed to all the institutions, organisations and commercial companies who contribute to the work of WFH. It is a healthy organisation that can critically appraise its own functions, and decide that some organs have outlived their usefulness. WFH did this when it accepted the recommendation from the Chairman of the European Advisory Board that the Board should be wound up, having achieved all that it set out to do. A most important resolution on AIDS was proposed and adopted by the General Assembly. AIDS was clearly a major topic of discussion at the Congress, and hardly a session passed without some reference to this problem.

On Wednesday evening we were privileged to be the guests of the City of Stockholm and the Stockholm County Council at a Reception in the City Hall. Built in the 1920's, this hall is truly magnificent and contains the breathtaking Gold Hall where the Nobel Prizewinners receive their awards. Here we sampled a traditional smorgasbord before the WFH presented a number of its awards to distinguished personalities in the world of haemophilia.

Back to work at lectures on Thursday, I attended a lively session on different views of the treatment of synovitis in haemophilia. Different approaches were discussed, including major surgery, arthroscopy and laser beam surgery. I found the video of an arthroscopic synovectomy quite fascinating.

Hypnosis in Haemophilia came next for me, a presentation by two hypnotherapists working in Colorado. Their patients show an apparently diminished incidence of bleeds as a result of hypnotherapy, and while I would treat their claims with some caution, I was sorry that such a large number of delegates left this session before the end. I feel it is very

important to keep an open mind on such topics.

In the Orthopaedic session in the afternoon, I enjoyed two very worthwhile contributions – one from Mr. Mike Smith (St. Thomas's Hospital, London) and the other from Dr. A. Aronstan, on the work he is doing at Lord Mayor Treloar College.

The final session that day on Inhibitors was largely beyond my understanding, and for me this is an area which requires attention in the future.

On Thursday evening, our Swedish hosts kindly organised house parties for all delegates, and this evening was much appreciated by all who attended.

The morning session on Friday comprised a miscellany of papers under the heading "Basic Studies – Various". As they were largely technical in nature, I took the opportunity to make a final tour of the posters and exhibitions. Attracted by a poster session display about TENS – Transcutaneous Electric Nerve Stimulation – I was able to attend a practical demonstration of this treatment, which it is now claimed not only relieves joint pain, but also reduces swelling and increases the range of movement in affected joints. Among the commercial displays I found a useful self-treatment training aid on the Hyland-Travenol stand. This is a set of flash cards with a Dennis the Menace-type cartoon character giving a demonstration of self-infusion. For anyone involved in patient education I think these are well worth seeing.

The afternoon session on Side Effects of Treatment attracted a packed hall, eager to hear the latest situation regarding AIDS. Drs. Aledort and Evatt, both of the USA, led an excellent symposium on the subject, and by 5.00 pm the Congress was over.

How would I summarise my first experience of a WFH Congress? I must first declare that I had no idea of what to expect from the Congress before I went. I viewed it as the top event in the world of haemophilia, and my expectations were certainly confirmed in terms of the amount of interest and medical research taking place in relation to haemophilia around the world.

I found it in the main a well-organised Congress. The Chairmen of the symposia performed sterling work in keeping to time schedules, and every minute of every day was usefully occupied. My only regret was that there was just not enough time in the day to get to know more delegates from other countries.

I confess to being slightly disappointed that many of the papers were so technical in nature, although I think it is an indication of the esteem in which WFH is held that so many professionals want to present their work at these Congresses.

I appreciate the value of WFH Congresses as a forcing ground to make those countries which are less advanced in their approach to haemophilia care more aware of what can be done.

I enjoyed and appreciated the experience very much, and I take this opportunity to thank the Executive Committee and the Society for enabling me to attend.

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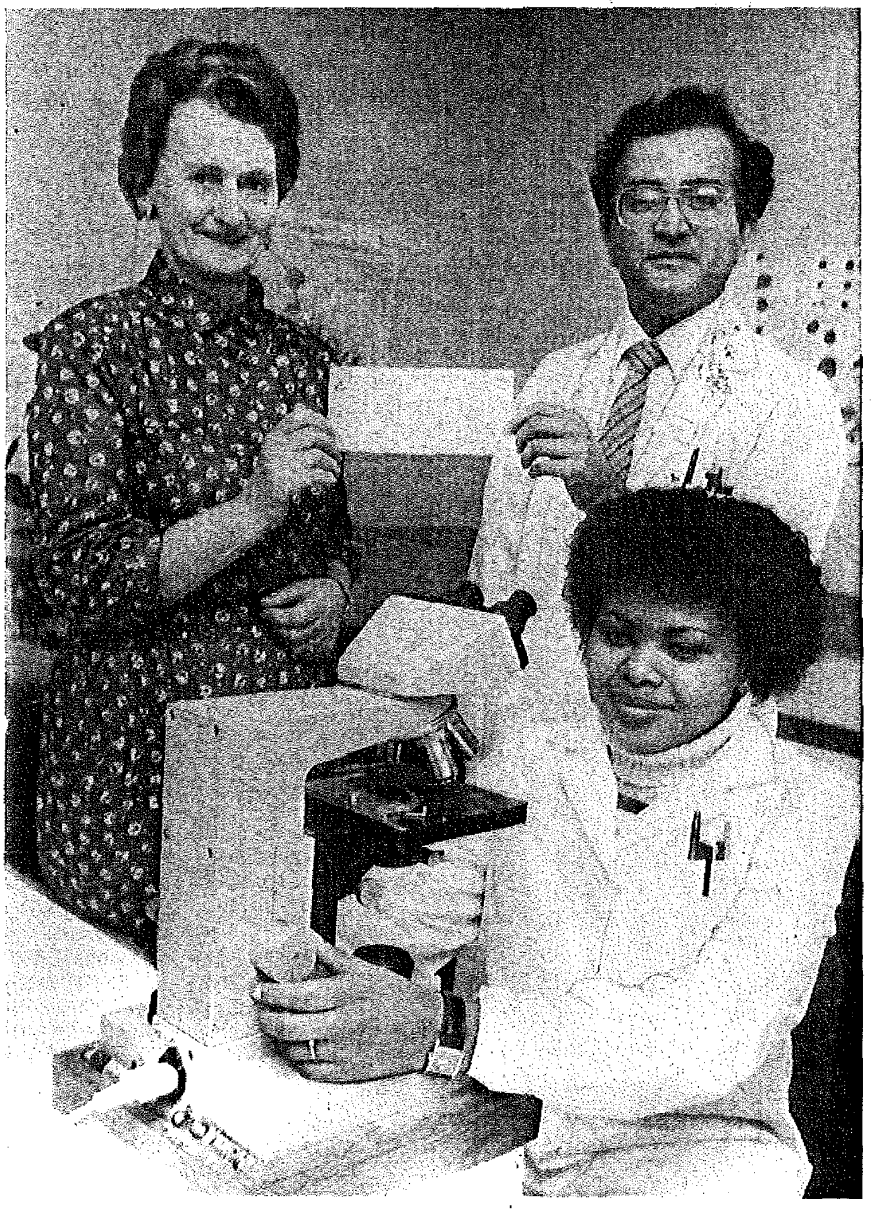
THE ACQUIRED IMMUNE DEFICIENCY SYNDROME (AIDS)

Dr. Anthony J. Pinching, BM, BCh, MA, DPhil, MRCP,
Senior Lecturer and Consultant Immunologist, St. Mary's Hospital Medical School, London W2

Certain types of infection (called opportunist infections) and some rare tumours are only seen in people whose immune system is defective. Such patients may be born without a part of their immune system (congenital immunodeficiency), whilst in others it is a result of a specific disease or treatment (e.g. in preventing kidney transplant rejection). In 1979-80, it was noticed that a particular pattern of such opportunist infections and tumours was occurring in a group of people who did not have any previously known cause for defective cellular immunity; this new disorder became known as the acquired immune deficiency syndrome (AIDS). Although it was originally described in male homosexuals, it has now been noted in intravenous drug abusers, Haitians and haemophiliacs, as well as in female sexual partners of AIDS patients.

The pattern of this epidemic has suggested that AIDS may be due to an infectious agent, transmitted by intimate contact or blood product inoculation, in a way reminiscent of Hepatitis B virus. While there are many other suggested causes, this one currently seems the most likely. The agent is probably a virus but it has not so far been identified; there are therefore no specific tests for it. A particular problem is that there appears to be quite a long period (months or years) between exposure to the causative



Mrs Irene Clinton (Chairman of our Northern Group) presenting a cheque for £3,000 on behalf of the Haemophilia Society to Dr Parapia, Director of the Haemophilia Centre at Bradford Royal Infirmary. The money will support research work being undertaken by Angela Kasote (front).

agent and the person becoming ill; during this time he/she may be infectious. One can try to detect evidence of the immune deficiency caused by this agent before symptoms develop. Unfortunately, the tests available, such as those of so-called T helper and T suppressor lymphocyte numbers and ratios, are extremely non-specific and may be affected by many other factors. Our own studies on male homosexuals in London have served to emphasise this point. The current position is that no immunological tests have yet been shown to be reliable as a way of screening at-risk individuals.

Over 2,000 cases of AIDS have been reported in the USA, while in Europe there have been about 180, and in the UK 24. However, the disease carries a high mortality, especially in cases with opportunist infections; while some of the infections and tumours are treatable, no cure has yet been found for the underlying defect of immunity. For this reason, despite the relatively small number of cases, the syndrome is rightly being viewed with some concern. In the present state of knowledge, a major objective must be to try to reduce the risk of transmitting the disease further.

How does this affect haemophiliacs? AIDS has affected roughly 1 in 1,000 haemophiliacs in the USA, and two patients in the UK. The immediate source of infection in such patients is thought to have been Factor VIII concentrate, derived as it is from thousands of donors. On the other hand, this new and to some extent theoretical hazard of using concentrates has to be set against the enormous benefits of such concentrates in haemophiliacs, especially for home therapy. As in any other medical setting, the risk of treatment has to be balanced against the dangers of the disease itself. Factor VIII concentrate from the USA may be the most likely to contain the AIDS agent; however, the risk is probably small and no source can be regarded as completely free from risk. Furthermore the USA is the only country capable of providing the quantity of Factor VIII currently needed by UK haemophiliacs. US producers of Factor VIII concentrates have already acted to reduce the risk of transmitting such an agent. The present balance of opinion among haemophilia centre directors in the UK therefore is that imported Factor VIII concentrate should continue to be used for those selected patients already receiving it: i.e. severely affected haemophiliacs with frequent bleeds, and excluding children and those with mild disease. The source of Factor VIII concentrates will need to be kept under constant review, as will blood donor policy, both by the medical profession and the relevant industrial concerns, to minimise or eliminate the risks.

AIDS is an epidemic new form of cellular immune deficiency; it appears to be caused by a viral agent transmitted by intimate contact or blood product inoculation. Currently the disease is resistant to treatment; no specific tests for it or the causative agent exist. It has continuing implications for many members of the population, but these need to be kept in a proper perspective. Further items on AIDS and other matters of topical concern will appear in our new pamphlet series HAEMOFACT which will appear at appropriate but irregular intervals.

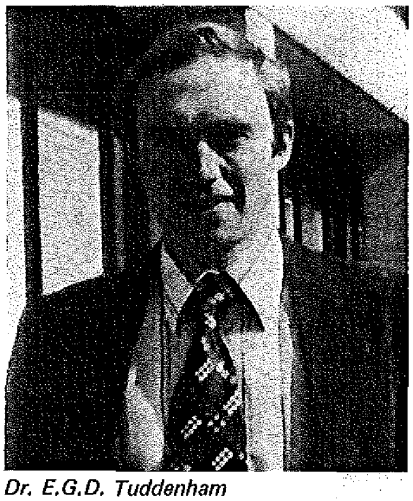
HAEMOPHILIA HOME THERAPY

A limited number of these books are available at a cost of £8 to Society **members** (this includes postage and packaging). This book will be of special interest to Centre staff and others involved in the professional care of people with haemophilia. Order your copy from 16 Trinity Street, London SE1 1DE.

INNOVATIVE ALTERNATIVES TO HUMAN FACTOR VIII

By E. G. D. Tuddenham

Let me begin by describing a process for you, one that is very familiar and carried out in all developed countries. A heterogenous natural product in liquid form derived from many individual citizens is pooled centrally. Various substances are added to it and precipitation occurs. A sediment is formed which is washed, treated further and finally dried. The end product is then widely distributed for use in the community. Now what process do you think that might be? Production of factor VIII concentrate?



Dr. E.G.D. Tuddenham

No, actually I was describing a sewage treatment plant. The basic principles of sludge separation closely resemble those of fractional precipitation as used in large-scale plasma fractionation and they are scientifically and technically at about the same level of development! I submit to you that the technology of factor VIII concentrate production has got stuck at a point reached by about 1970.

I don't know whether to laugh or cry when I hear people talk about "high purity concentrate", with a specific activity of 3 units/mg. Even to **use** the word purity in this context is an unwarranted exaggeration. Pure factor VIII has a specific activity not of 10 units/mg nor 100 units/mg nor yet 1,000 units/mg but at least 5,000 units/mg. That is what our fractionators should be aiming at! Instead of which you hear them congratulating themselves when they achieve a so-called potency of 7 units/mg.

What you actually see in the prettily packed bottle with the fancy label is a huge cake of high molecular weight proteins (fibrinogen, fibronectin, IgM, viruses etc.) very, very lightly contaminated with the protein the patients need — factor VIII coagulant. On one of the exhibition stands outside the hall I saw a polyacrylamide gel on which were analysed the proteins from 9 factor VIII preparations. Dozens of protein bands can be distinguished but of factor VIII there is not a trace. Is there any other pharmaceutical that we are prepared to inject into human veins at a purity of less than 0.1%? If there is I don't know of it — though factor IX concentrate — so called — comes a pretty close second at 1% purity.

Now who is to blame for this deplorable situation the results of which are being discussed in the seminars on hepatitis and AIDS? Frankly, we are all in this together, doctors, patients, national blood centres, commercial fractionators. We must recognise the deficiency and take drastic steps to bring coagulation factor purification into the 1980's. I do not consider that heating and irradiating blood products to remove things that shouldn't be there in the first place is anything other than a temporary measure of desperation.

Change is urgent but we are up against entrenched methods and obstructive legislation in some countries. As an example, consider the polyelectrolyte fractionation process just reviewed by Dr. Johnson for us in the previous presentation. This was described in 1977 and provides a method to make much higher purity factor VIII, factor IX, albumin, IgG and fibrinogen with probable removal of all or most viruses from the major coagulation factor fractionations. Was this avidly and widely taken up and determinedly developed worldwide? By no means!

Ultimately it's a question of having the will to persevere with developments to bring improvements into large-scale production, despite the inevitable teething problems of any new method.

Looking beyond improved ion or hydrophobic exchange resins, we are now in the era of monoclonal antibodies. This technology introduced by Milstein in 1976 offers us the chance to use nature's own unique specific system developed over millions of years to recognise and bind protein antigens out of mixtures. I refer of course, to the immune system which functions to recognise foreign proteins and produces immunoglobulin molecules that precisely and avidly latch onto exactly defined areas of proteins. One can induce immune cells to react against an injection of human factor IX in a mouse spleen. Then one can fuse them with tumour cells and select the hybrids that make antibody to human factor IX. One then has a clone of cells that grow indefinitely and produce limitless quantities of a pure factor IX binding antibody.

In my laboratory, in the past three years (working with Dr. Goodall in the

Immunology department of the Royal Free Hospital) we have created clones of immortal mouse cells that make specific antibodies against factor IX, von Willebrand's factor and against the factor VIII coagulant protein (VIII:C).

We have used these to purify to complete purity factor IX and factor VIII:C. For example we have a 3 ml column of agarose (about a teaspoonful) to which is bound monoclonal antibody to VIII:C and this tiny column has a capacity for about 50,000 units of VIII:C. It completely takes up all the VIII:C that we have ever applied to it, and after washing we recover the VIII:C at 100% purity in 90 to 100% yield. I believe that monoclonal antibodies offer us the tools to make our factor concentrates deserving the epithet – "high purity".

Turning from human plasma fractionation, I want to focus your thoughts on the following facts provided to me by the U.K. Meat and Livestock Commission. In 1982 the following animals were slaughtered for meat in the British abbatoirs:– 15 million pigs, 14 million sheep and 3.6 million cattle. Note that they were processed for meat and the vast majority of the blood was simply washed down the gutters. At a conservative estimate, that blood contained 633 thousand million units of VIII:C. Of course this huge potential source of factor VIII did not escape the attention of the pioneer workers at Oxford who in the 1950s and early 60s prepared large amounts of pig and cow factor VIII that was successfully used to cover many surgical procedures in haemophiliacs. However the drawbacks of these early concentrates in terms of allergic reactions and thrombocytopenia induction led to their general abandonment as human factor VIII concentrates became available. In the past 4 years a new start has been made on recovering a little of this massive national wastage of animal factors by the development at Speywood Laboratories of a new process based on polyelectrolytes. The seminar on Thursday is largely devoted to porcine factor VIII in its new form of Hyate C so I will not expand at length on this topic other than to say that there is absolutely no doubt that Hyate C is highly effective haemostatically in a proportion of inhibitor patients. Also that up to a half of inhibitor patients do **not** develop resistance so that long-term therapy is possible. I would suggest that we now can start thinking seriously about the possibility of using Hyate C in certain non-inhibitor factor VIII cases who would be at very high risk of developing hepatitis if given standard human concentrate. To date there are no reported cases of AIDS in pigs.

In a talk on innovative alternatives I feel I should mention the recent report in Haemostasis (Vol. 13: pp.78-82, 1983) from Japan of the use of a herbal mixture called Huang-lin-chieh-tu-tang, which when administered orally to a severe haemophiliac, elevated his factor VIII level from <1% to 41%. Also recently reported by Gralnick & Rick in the New England Journal of Medicine were several moderately severe cases of haemophilia A whose factor levels rose 3 or 4% after administration of Danazol, an attenuated androgen. These and other alternative therapies should certainly be given serious consideration at a time when we are all very seriously worried about the side effects of conventional treatment.

Finally what hopes are there of making factor VIII by recombinant DNA methods – genetic engineering is the popular phrase so often seen in the press these days.

Factor VIII:C is a gene product, a protein like other coagulation factors and although it resisted purification for longer than any other factor it is finally yielding up its secrets. In my laboratory we have concentrated factor VIII:C from several tons of plasma over 300,000 fold to make pure protein for sequence analysis. Other laboratories are engaged in this task and I venture to predict also that the full structure of the protein and of the gene will be known in the not too distant future.

My colleague Gordon Vehar of Genentech gave an excellent summary and introduction to the Bioengineering of blood derivatives in the lecture hall earlier in the week so I will only repeat his prediction that it could take 5 years to see bioengineered factor VIII free of all viruses and other contaminants on the market. We have to live in the meanwhile and that is the period in which the improved fractionation methods must operate from human and animal blood sources.

Finally to the wilder shores of speculation. What chance of gene therapy? I, follow this field as an outsider but I have been encouraged by experiments in which mammalian genes have been passaged between species and shown to function. A recent cover of Nature was graced by a picture of giant mice. This miracle was achieved by implanting cloned rat growth hormone gene, attached to a promoter sequence, into fertilised mouse ova. The mice that developed therefrom had largely elevated levels of rat growth hormone in their serum, and grew to an abnormally large size, thus proving integration and function of a mammalian gene after cloning from another species. One thinks of applying this to the human VIII:C gene. Could it be integrated into a fertilized cow's egg, and the resulting cow milked for millions of units of human VIII:C? We do not know which cell types make VIII:C but if available to biopsy and tissue culture one can envisage taking cells from an adult haemophiliac, genetically 'correcting' them by introduction of a cloned normal gene and returning to the patient. Let me finish with a quotation from Fitzgerald's poem The Rubaiyat of Omar Khayyam –

> "Ah love could'st thou and I with fate conspire,
> To seize this sorry scheme of things entire,
> Would we not shatter it to bits, and then
> Remould it closer to the heart's desire".

Abstract No. 37 given at W.F.H. Stockholm, 1983

★★★★★★★★★★★★★★★★★★★★★★

MANUFACTURERS' DISCOUNT SCHEME

Manufacturer: Austin Rover
Discount offered: Up to 17%.
On Models: All models.
Eligibility: MA & car allowance recipients NHS vehicle service beneficiaries.
Contact: Mr. Malcolm Jones, Mobility Dept., Austin Rover Group, Longbridge, Birmingham B31 2TB.

Manufacturer: Fiat.
Discount offered: 12% on all basic models 15% on all other models.
Eligibility: MA recipients, their parents, immediate family, guardians, etc.
Contact: Mr. M. Lee, Sales Director, Fiat Auto UK Ltd., Great West Road, Brentford, Middx. TW8 9DJ.

Manufacturer: Ford
Discount offered: 21% approx.
On Models: Escort Auto Saloon Estate with disability pack.
Eligibility: MA recipients and registered disabled persons.
Contact: Mr. Bob McAuley, Ford Motor Co. Ltd., Brentwood, Essex CM11 3BW.

Manufacturer: Renault.
Discount offered: Announcement in near future.
Contact: Mr. M. Mead, Fleet Sales Manager, Renault UK Ltd., Western Avenue, London W3 0RZ.

Manufacturer: Saab.
Discount offered: 14% excluding metallic paint & sun roof.
On Models: All models.
Eligibility: MA recipients and NHS vehicle beneficiaries.
Contact: Alison Jones, Public Relations, Saab (GB) Ltd., Fieldhouse Lane, Marlow, Bucks. SL7 1LY.

Manufacturer: Talbot.
Discount offered: Negotiable with your dealer for discount recommended by Peugeot Talbot.
On Models: All models.
Eligibility: MA recipients & NHS vehicle beneficiaries.
Contact: Mr. R. G. Oakley, Manager Leasing & Rentals, Peugeot Talbot, PO Box 46, London Road, Ryton-on-Dunsmore, Coventry CV8 3DZ.

Manufacturer: Vauxhall Opel.
Discount offered: 14% on Chevette; 17% on Astra, Cavalier; Negotiable on Manta.
Eligibility: MA recipients & NHS vehicle beneficiaries.
Contact: Graham Pitney, Fleet Sales, Vauxhall Motors Ltd., PO Box 3, Luton, Beds. LU2 0SY.

Manufacturer: Volkswagen.
Discount offered: Up to 12½% negotiable with dealer.
On Models: All models.
Eligibility: MA recipients & NHS vehicle beneficiaries.
Contact: Mrs. M. McShane, Sec. to Sales Manager, Volkswagen (GB) Ltd., Yeomans Drive, Blakelands, Milton Keynes, Bucks.

Manufacturer: Volvo.
Discount offered: 15%.
On Models: All models.
Eligibility: Any disabled person registered with local authority, employment service agency, etc., MA recipients. Not necessarily a driver.

Contact: Daphne Why, Customer Information Administrator, Volvo Concessionaires Ltd., Lancaster Road, Cressex Estate, High Wycombe, Bucks. HP12 3QE.

The foregoing figures are the discounts offered on the basic price of each vehicle with appropriate adjustment to VAT, but Car Tax in each case remains as listed on the sales literature.

These discounts are offered by the manufacturers themselves, but some dealers may be reluctant to provide vehicles on these terms, as it is the dealers' profit margins and subsequently the salesman's commission, that bear the brunt of these price reductions. Shop around and if at first you don't succeed, try, try again!

Positively no trade-in can be considered on these terms at any time.

Special credit terms are available from some manufacturers and details can be obtained from the contacts listed.

HOW ABOUT . . . A DIMINISHING TEA PARTY!!

This is how it works . . . a member of the Society starts the scheme by inviting twenty guests for coffee/tea – in the morning, afternoon or evening. Each guest is then invited to donate, say, £1 to the Haemophilia Society, and to then invite six of their own friends to their homes for tea/coffee, each guest contributing £1. Each of these invited five guests – so the numbers DIMINISH – but the money raised INCREASES like this:–

Round 1 1 hostess with 20 guests – raises £ 20
Round 2 20 hostesses with 6 guests – raises £ 120
Round 3 120 hostesses with 5 guests – raises £ 600
Round 4 600 hostesses with 4 guests – raises £ 2,400
Round 5 2400 hostesses with 3 guests – raises £ 7,200
Round 6 7200 hostesses with 2 guests – raises £14,400.

A total of £24,740.

HOW ABOUT IT THEN??

(With thanks to the Haemophilia Society of Victoria, Australia.)

THE RESEARCH APPEAL

You will be interested to learn that the Research Appeal Fund is ALMOST THERE – at the time of writing a total of some £247,000 has been raised – so if you have a stocking collection or a collecting box, send it in and enable us to round off the appeal this year!!

Needless to say, we are all deeply grateful to everyone who has contributed to the appeal and send our heartfelt thanks.

IF YOU ARE OVER 65 . . . and wish to claim Mobility Allowance: the answer is that you cannot do so. The Co-ordinator recently received a letter asking about this. Unfortunately the letter was mislaid and he has been unable to answer directly. It is hoped that the writer of that letter reads this piece. The Co-ordinator sends his apologies.

THE LONDON MARATHON

A BIG THANK YOU to everyone who collected sponsorship for PETER STEVENS in the London Marathon. Thanks to your generosity a total of £2,916.83 was raised. We were most impressed by the number of people who telephoned the answering machine to find out the result – the digital counter stops at 99 calls, so we have no real way of knowing just how many people were keen to discover the result . . .

Our photograph shows DAVID ODDY at the end of the Marathon. He ran for the benefit of the Society in company with his brother NIGEL. They took 4½ and 3½ hours respectively to finish . . . and raised the splendid sum of £333.00 for the Research Appeal. Our thanks to them and a number of other thoughtful people who have taken part in sponsored events to benefit the Society.

The **ANNUAL DRAW** made an all-time record of just under **£4,000** – our warm thanks to all who sold tickets. **NEXT YEAR there will be a special prize for the member selling most tickets – and someone sold over 150 books this year!!**

OVER 16 AND STILL AT SCHOOL?

If you have haemophilia and have stayed on at school to improve your eventual prospects of employment you may very well be entitled to Supplementary Benefit IN YOUR OWN RIGHT. That is to say, it does not depend in any way on your parents' income; otherwise normal DHSS Regulations apply. SNAGS? Your parents will lose Child Benefit payable in respect of you, but as a family you should not be worse off. Assuming you are entitled and have no other income (eg educational grant) the payment could be worth £28+ per week.

WORLD FEDERATION OF HEMOPHILIA PUBLICATIONS

Got your PASSPORT yet?

PASSPORT – the new Guide for Travelling Haemophiliacs is now available from the Society's offices. This is published by the World Federation of Haemophelia and prepared by ALPHA Therapeutics Corporation.

PUBLISHED BY THE HAEMOPHILIA SOCIETY

Series:
INTRODUCTION TO HAEMOPHILIA

- Notes for Parents
- Notes for Teachers
- Notes for Health Visitors
- Notes for Social Workers
- Notes for Careers/Employment Services

SURVEY REPORT (Published 1977)

THE HISTORY OF HAEMOPHILIA

THE BULLETIN – FREE TO ALL MEMBERS

There are also sundry and occasional reports: always feel free to get in touch with the Society's office if there is any subject you wish to know about.

HAEMOPHILIA HOME THERAPY

by Dr. Peter Jones

A **limited** number are available at a cost of £8.00 (incl. p & p) to Society members. This book will be of special interest to Centre staff and others involved in the professional care of haemophiliacs. Please order your copy from the Co-ordinator, at 16 Trinity Street, London SE1 1DE.

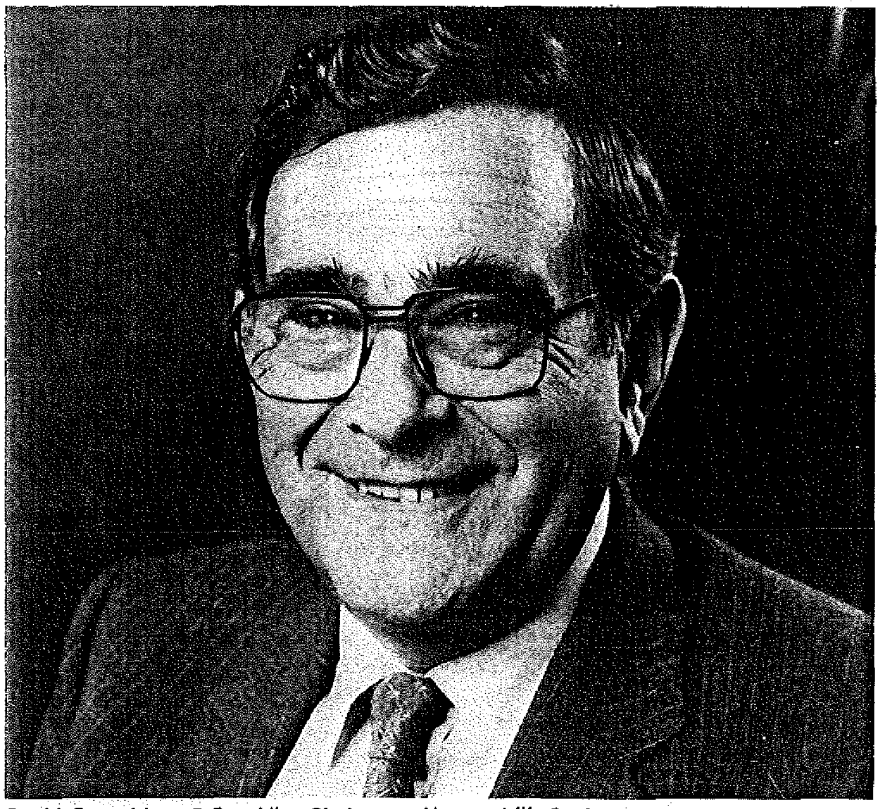
YOUR EXECUTIVE

David Rosenblatt, BSc, Vice-Chairman of the Haemophilia Society: Educated at St. Marylebone Grammar School and King's College, University of London, and now Managing Director of a registered insurance brokerage. David and his wife Sylvia have one son, aged 29, who has Christmas disease (Haemophilia B). Their son is now married and is a practising solicitor in a two-man partnership in Watford, Herts.

David Rosenblatt became interested in Society work, partly as a result of his son attending the Royal Free Hospital, having previously been a patient of the Hospital for Sick Children, Great Ormond Street. It was at the Royal Free that David was persuaded to stand for election to the Executive Committee by Mrs. Peggy Britten!

In his work for the Society David has been especially interested in fund-raising, life and travel insurance for people with haemophilia, in addition to the normal work of a member of the Executive Committee. During the 15 years in which he has served on the Executive Committee and Council, David has represented the Society at WFH meetings in Moscow, Bonn, Tel Aviv and attended the 1976 Congress in London.

Interests: Gardening, music, soccer and the welfare of people with haemophilia.



David Rosenblatt, B.Sc., Vice Chairman, Haemophilia Society

Howard Abrahams

Basic reason for wishing to become involved with the Society is due to my strong desire in early seventies to effectively put back into "system" what I had taken out, etc. (bit of a moral obligation really).

My father was a former Executive member in early fifties so our family had connections with Society. I also felt that my financial experience and involvement as a practising Chartered Accountant would be of benefit to the financial well being and running of the Society.

Finally as a patient at the Royal Free Hospital, I came into contact with Petty Britten who was a great encouragement to me joining the "team".

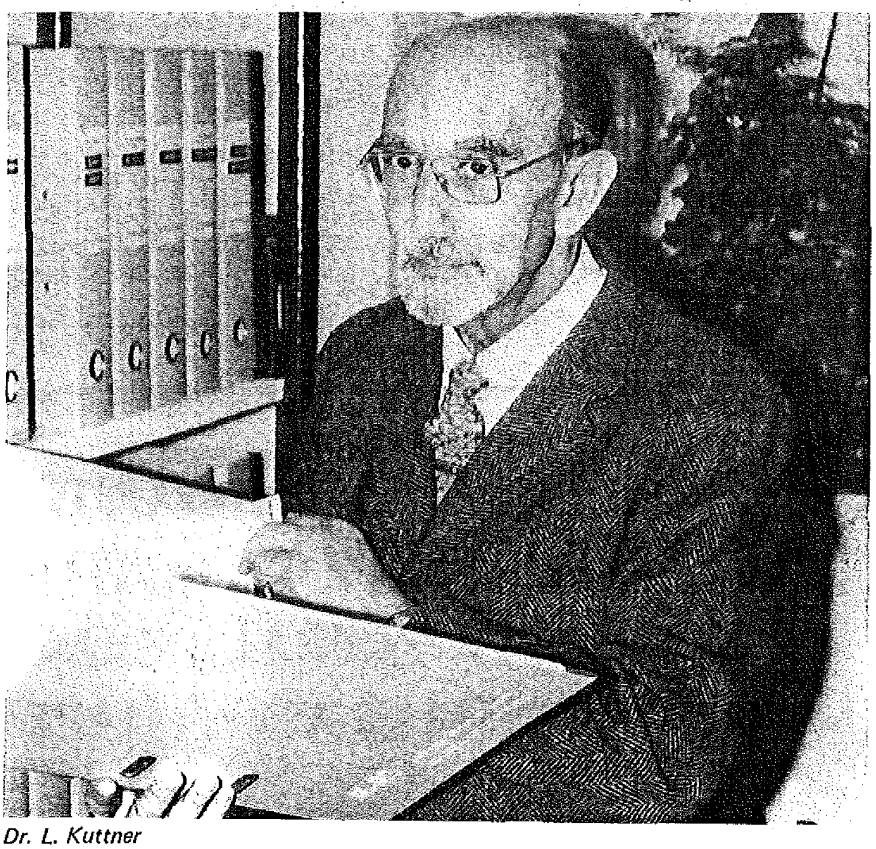
Howard Abrahams

There has been haemophilia in my wife's family for many generations and when my son was 15 months of age he showed signs of haemophilia. That was more than 31 years ago and ever since my wife and I have been associated with the Society.

I have been Chairman of the North Eastern Group (then Leeds) for 5 years and of the Southern Group for 3 years.

For over 10 years I was Group Liaison Officer of the Society and have been a member of the Executive Committee for several more years.

Dr. L. Kuttner



Dr. L. Kuttner

SOCIETY CHRISTMAS CARDS

We are happy to announce that Society CHRISTMAS CARDS are available this year. Stocks are held by the following LOCAL GROUPS:–

There are ten designs available – and they range from the conventional to the zany. Prices on the other hand range from 67p to 70p for a pack of six cards (all the same design in each pack). **PLEASE CONTACT your nearest local Group for supplies of those cards.** We only hold very limited stocks at Trinity Street.

Cambridge Group
Contact: Mrs. F. Wetherell
47 Manor Place
Cambridge CB1 1LJ

Leicester & Rutland
Contact: Mr. B. T. Lewis
335 Butterwick Drive
Beaumont, Leys
Leicester LE4 0UH

Lewisham
Contact: Mr. A. Weir
147 Wellmeadow Road
Catford
London SE6 1HP

Northern Ireland
Contact: Mr. S. Graham
15 Keel Park
Moneyreagh
Co. Down BT23 6DE

North Wales
Contact: Mrs. C. Holliday
54 Bastion Gardens
Prestatyn
Clwyd LL19 7LU

Oxford Group
Contact: Mrs. B. Weir
The Old Stables
Market Square
Lechlade, Glos. GL7 3AB

Scottish Group
Contact: Mrs. S. Cowe
106 Houstoun Gardens
Millburn Park
Uphall
West Lothian EH52 5SH

Southern Group
Contact: Mrs. E. Burrows
12 Tennyson Road
Wimborne
Dorset BH21 1NT

South Wales
Contact: Mrs. D. Lewis
'Dolwerdd'
82 Parc-y-Coed
Creigiau
Mid Glam CF4 8LZ

AT THE ROYAL FREE HOSPITAL

HYDROTHERAPY & PHYSIO/GYM
Restarted on Monday, September 19th and thereafter **Every Monday** 6–8 pm
HAEMOPHILIA EXERCISE GROUP
includes . . .

Pool Exercises
Gym Exercises
Team Exercises
Great Fun!!

Open to patients from other Centres – all you need to do is bring a letter from your own Centre and treat yourself beforehand if appropriate.

To book contact: Karen Beeton
Physio Department
Royal Free Hospital
Pond Street
Hampstead NW3
Tel. 794 0500 bleep 548

Views expressed in the Bulletin are not necessarily those of the Haemophilia Society.

A CARAVAN HOLIDAY IN 1984?

When thinking of your 1984 holiday, don't forget that the Society have holiday caravans for members throughout the country. The rates are most attractive (and much cheaper than commercially available vans). The people to contact are:

Mid-Wales: Situated at Borth
Contact person: Mrs. K. Instone
21 Cheswick Way
Cheswick Green
Solihull
West Midlands B90 4EZ
Tel. No.: 056 46 2975

Bognor Regis:
Contact person: Mrs. J. Copping
39 Hawes Lane
West Wickham
Kent BR4 0DA
Tel. No.: 01-777 7746

Clacton-on-Sea:
Contact person: Mr. Peter Frost
22 Winston Way
Halstead
Essex CO9 2TH
Tel. No.: 0787 475109

Lancaster: Situated at Caton
Contact person: Mrs. A. Marks
10 Alston Road
Whelley
Wigan WN2 1AU
Tel. No.: Wigan 38259

ENCLOSED with this edition of THE BULLETIN you will find an offer for beautiful home-knitted sweaters in traditional Fair Isle and Icelandic patterns. They are knitted by hand on the Island of Hoy in the Orkney Islands. The offer is limited to members of the Haemophilia Society and their friends; meantime, 10% commission is payable to the Society by the knitters.

Also enclosed is a list of winners in the GRAND DRAW. Thanks to all those who sold tickets and congratulations to the winners. This year the Draw took more money than ever before, almost breaking the £4,000 barrier in income!

THE CHAIRMAN, EXECUTIVE COMMITTEE AND STAFF OF THE HAEMOPHILIA SOCIETY WISH ALL READERS A MERRY CHRISTMAS AND A HAPPY NEW YEAR