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The Bulletin

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

Member of the World Federation of Hemophilia
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THE HAEMOPHILIA SOCIETY
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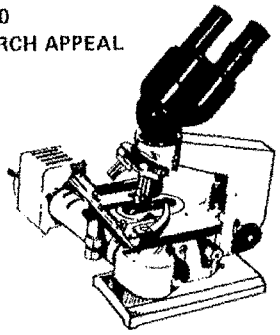
£250,000 RESEARCH APPEAL



On the 1st May 1979 our £250,000 Research Appeal was officially launched as a "MAYDAY" appeal and, although getting off to a slow start impetus has increased so that in under one year over £40,000 has been raised.

About 75% of this sum has come from our Groups so ably led by their local committees, whose hard work and enthusiasm are a marvellous example to us all. The variety of events organised and sums raised are almost unbelievable!

Most Groups have made substantial donations and we would especially mention Northern Ireland £10,000 — Merseyside £2,700 — Jersey £1,900 — Great Ormond Street £2,200 — Northampton £2,000 — North West (Manchester) £2,500 — South Wales £2,500 — Northern (Newcastle) £2,700. Other Groups, some quite recently established, have played their part and details are given in this Bulletin.

Individual members have also sent generous contributions and organised fund raising functions and a stream of letters regularly leaves our office addressed to companies, trusts etc., seeking help and support.

Grateful thanks go to all those concerned in raising the largest amount ever achieved by the Society in such a short time. This money has been raised by people not only for hospitals, which they themselves attend, but also for hospitals in other areas, where, in the judgement of the Society's Council and Medical Advisory Panel, the projects being undertaken will be of most benefit and value to haemophiliacs and their families.

One such project is that on "Pre-natal Diagnosis of Haemophilia" being carried on jointly by Professor Arthur Bloom and Dr. Ian Peake (who have been awarded the French Haemophilia Association's International Prize for their work on a new assay method for Factor VIII) at the University of Wales, Cardiff, and Dr. Reuben Mibashan and Mr. Charles Rodeck of King's College Hospital, London.

In the past some women, who were likely to be carriers, when told that their unborn child was male, chose to have their pregnancy terminated, rather than risk the birth of a haemophiliac son. This, of course, meant that some normal babies were being aborted.

Now, thanks to new techniques developed in co-operation by the two hospitals mentioned, it is possible to determine whether or not a male foetus has haemophilia.

This is a tremendous step forward and can fairly be described as "life saving" work. It is an advance which could transform the lives of many carriers, some of whom have refrained from having children, or even from marrying, because of the possibility of having a haemophilic son. It will obviously be of interest to all haemophiliacs with a daughter, a sister, a cousin, or other female relative, who may be a carrier.

Because of the importance of this work the Society has responded in a positive way to requests for financial help from the two hospitals concerned. £10,000 has been granted to the University Hospital of Wales to pay for a Research Technician for 2 years and £12,606 to King's College Hospital for Research Staff and new Coagulometer Instruments.

In our Bulletin No. 3 in 1979 we anticipated that "further calls will probably be made upon the Society's Research Fund in the future to finance projects in this comparatively new and exciting field".

These calls may have arrived more quickly than expected but we feel sure that all in the Society will agree that the grants made are fully justified and will bring the day nearer, when there is a national pre-natal diagnosis service available to all who need it.

We hope that you will continue to give your support, in every way possible, to our Research Appeal.

CONTENTS

VICKI STOPFORD

Readers of the Bulletin will be familiar with the features of Mrs. Victoria Stopford, the Society's Research Social Worker, for her photograph appeared regularly to mark her column, "Your Problems".

In her replies to letters from members, Vicki revealed a wide-ranging acquaintance with all aspects of haemophilia and the needs of our members. She dealt with the severely practical matters such as caravan holidays and the benefits available from

INNOVATIONS FOR HAEMOPHILIACS

Dr. Charles Forbes is confident that improved care for difficult patients should lead to less arthropathy and analgesic misuse.

In the past 15 years there has been a dramatic improvement in the management of patients with haemophilia. This is due directly to a method of producing a simple cheap concentrate of factor VIII by the method of cryoprecipitation of human plasma.

For the first time it seemed possible that enough human material could be provided for routine and emergency use by using a high percentage of standard blood donations. The method is simple, relatively reliable and does not require sophisticated apparatus. It is therefore ideally suited for Regional Blood Transfusion Services and up to 40 per cent of all donations in many areas are used for this purpose. It is clear that this was the turning point in haemophilia care.

Many sophisticated concentrates are now being prepared with cryoprecipitate as the starting material. While they have many advantages they are extremely expensive and provision of adequate supplies remains in the political, rather than the medical, arena.

Availability of such preparations has led to more aggressive approaches to therapy, the most important of which is self-administration of the concentrate by the patient at the time of injury or at the start of a bleed (see figure). There is now little doubt of the major changes this has produced in the life-style of the severely affected patient.

An extension of this procedure is the routine administration of material to protect against bleeding. This is extremely costly, consumes large amounts of material and can only be recommended in selected patients at the moment, but should be the ideal goal for all patients.

There seems to be no evidence of serious side effects from this increased exposure to plasma products, in particular no increased incidence of factor VIII inhibitors or of hepatitis B carriers.

There can be no doubt about their benefits, with normalisation of social life, reduction in bleeding episodes and better attendance at school and work. The ultimate objective will be reduction of crippling from recurrent joint bleeds and normalisation of the life span of the patients – neither of which have been achieved as yet.

The most serious complication in the routine management of the haemophiliac is the development of an antibody to factor VIII in about 6 per cent of patients. This does not change the baseline level of the factor or increase the bleeding tendency, but does make treatment ineffective.

As yet no clear method of management is fully agreed on. Factor VIII – inhibitor bypassing activity (FEIBA) is widely used, but doubts exist about the theoretical basis as well as its place in management.

Products which contain activated factor IX may also be useful and recently concentrates of porcine factor VIII, treated to remove the platelet aggregating activity of the high molecular weight carrier protein, have been produced (Hyate) but these have not yet had sufficient clinical evaluation to assess their worth. There seems little evidence that long term immunosuppression is of value.

In a small number of very mild patients with reasonably high baseline levels of factor VIII, it is posible temporarily to boost the level to one which is haemostatic by the infusion of des amino D-arginine vasopressin which probably acts by stimulating release of coagulant activity from storage sites.

This overcomes the problem of using plasma concentrates in this small group of patients who seem to have a surprisingly high incidence of serum hepatitis after such exposure – presumably a reflection of a low level of immunity to the virus due to infrequent previous exposure.

Haemophilia Centre Handbook
Notes for Doctors and Nurses involved in the Care of Patients with Congenital Coagulation Disorders
by Jennifer Voke, Colin Madgwick & Katharine Dormandy
published by Immuno Ltd.
price £3.00
available from the Royal Free Hospital Haemophilia Centre
not available from bookshops

This inexpensive manual will prove valuable as an orderly source of facts for quick reference and as a teaching aid. It was written for the staff of the Royal Free Hospital Haemophilia Centre but is now available for anyone involved in the care of patients with inherited bleeding disorders. The book is dedicated to the memory of Dr. Katharine Dormandy who died while the manuscript was in the final stages of preparation.

Fibrinolytic inhibitors such as Aminocaproic acid (EACA) and Cyclokapron (AMCA) are of limited value and should be reserved specifically as an adjunct to concentrate in control of bleeding following dental extraction. There is no proof that they have any place in management of haemarthrosis or in patients with inhibitors.

Relief of pain from degenerative joint disease remains a problem for every patient and clinician in the haemophilia centre. One must temper humanity with the dangers of drug dependance and every centre has four or five such patients who have become addicted.

The drugs usually involved are dihydrocodeine (DF 118), dipipanone, and occasionally pethidine. These patients often have other evidence of personality disorders and are extremely difficult to manage. Hopefully with better treatment and orthopaedic care in the future we shall see less arthropathy and hence remove the trigger for analgesic misuse.

As medical treatment has improved the physical care of the haemophiliac, we have become more aware of the social and psychological stresses and strains in the patients as well as the families. These are currently under investigation and hopefully some will be amenable to counselling, both genetic and social, but this equally requires the motivation of the person involved and the will of society. Perhaps the most important force for good in the future is a powerful, motivated and active National Haemophilia Society.

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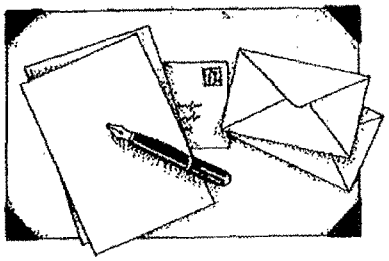
Charles Forbes is senior lecturer in medicine at the Royal Infirmary, Glasgow.

Points of View



From Mr. N. Wade,
40 Park Place,
Margate,
Kent.

Dear Sir,

I am writing to ask you if you would mention this letter in your next Bulletin. My son, Gary, went to watch Gillingham Football Club, which is quite a long way from Margate. While he was watching the match he got a severe bleed in the leg and told a Policeman, who was standing in front of him. The Policeman and his friend carried my son out of the ground, got him an ambulance and went with him to Margate Hospital (some 50 miles away) where Gary was treated. When they got back to Gillingham, he phoned me at Margate to ask how Gary was. I think this was so kind, that I cannot thank the Gillingham Police enough. I hope you can print this so that I can send them a copy.