The Bulletin

1989 — No. 3

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

Member of the World Federation of Hemophilia
Registered Charity number: 288260

The Haemophilia Society
123 Westminster Bridge Road
London SE1 7HR
Telephone: 01-928 2020

THE USA, HIV & VISAS

THE HAEMOPHILIA SOCIETY has protested in the strongest possible terms to the U.S. Embassy in London, and to Congressmen in the USA, following the implementation of an effective ban on anyone who is HIV positive entering the USA.

Despite consistent calls from the World Health Organisation and Council of Europe for unrestricted travel for people with HIV/AIDS, many countries still have immigration restrictions. These entry qualifications are mainly for individuals applying for work permits and are mainly from countries in the Middle East.

However, certain countries also pursue a policy of entry restrictions for people intending to visit their country for tourist purposes. Ironically, the United States is one such country which discriminates against the tourist with HIV infection. According to immigration policy, the person with HIV/AIDS can only gain entry into the United States as long as a waiver has been granted. These waivers can only be awarded if people are visiting the United States:—

- > to visit close relatives
- > to conduct short term business
- > to attend a conference or seek expert medical advice

Naturally, the Society is appalled at this situation. We have made our feelings clear to the U.S. Embassy in London. They, in turn, have agreed to contact Washington to have the policy reconsidered.

STOP PRESS

As we go to press it is reported that the US Embassy IS granting four week Visas to *children with haemophilia* who are going to the USA on holiday.

The Embassy will be able to advise you on the correct procedure to follow.

IF YOU ARE HIV POSITIVE YOU MUST HAVE THIS WAIVER.

The situation, as it stands, means that people with HIV cannot travel to the U.S. without consent, which may or may not be granted. This is of course a ridiculous situation, primarily for the reason that there is no way an immigration official can know a person is infected just by looking at him. Thousands upon thousands of people with HIV travel to the U.S. yearly, unbeknown to the immigration department. Problems can arise, however, if a person is stopped and quizzed over HIV.

Many people have decided to ignore the ruling and continue to travel back and forth without any problems, but it must be pointed out that recently a man with AIDS, identified by carrying AZT, was imprisoned overnight and sent back to London.

It is clearly advisable not to travel and face the prospect of being 'turned around', detained in custody and sent back on the next available flight – with no financial redress.

Hopefully, the situation will improve. If you require further information you can call the U.S. Embassy directly and speak to David Rollman (GRO-C). He is happy to receive any calls concerning immigration, haemophilia and HIV.

If you would like information on entry requirements into other countries, in connection with HIV, please contact the national office.

NHS Review

Are you for it or 'agin' it?

Well, to put it simply we are 'against it in the strongest possible terms – and we have informed Kenneth Clarke of that fact. We are deeply concerned about how it might affect the future of haemophilia care throughout the UK.

Copies of our initial response are available from the national office – a stamped addressed envelope would help. You should also contact your local Group about responses on a local level – see elsewhere for the address of your local contact person!

CONTENTS

Not Relevant

POVERTY AND DISABILITY: The Chance to Break the Link

Jonathan Cooper

As the last Update mentioned, 1989 is a vital year for people with disabilities in receipt of benefits. In order to review the welfare system, the Government has commissioned the Office of Population Censuses and Surveys (OPCS) to produce information on the level of disability and degree of hardship of affected people, throughout the United Kingdom.

All the reports have now been published and have shown some quite startling results. Of key interest is that there is an estimated 6.2 million disabled adults in Great Britain – the majority of whom are over pensionable age, more than double the previous official figure. These include some 360,000 disabled children.

Disturbing aspects of the reports have shown that the bulk of people with disabilities are living in or are on the margins of poverty. The average income of disabled adults, 75% of whom rely on state benefits as their main source of income, is £65.20 per week. From the findings of the OPCS reports it has been estimated that, once the extra costs of disability are taken into account, disabled non-pensioners have, on average, incomes which are £51-60 per week less than non-disabled people. Put in more universal terms, the total income deficiency of disabled people, compared to the non-disabled population, is nearly £8 billion per year.

Armed with this information the Government is in a unique position to reassess policy towards people with a disability. As the Conservative party declared in its 1979 manifesto,

> "our aim is to introduce a coherent system of cash benefits to meet the costs of disability so that more disabled people can support themselves and lead normal lives. We shall work towards this as swiftly as the strength of the economy allows."

The Government has concentrated over the last ten years on consolidating its resources. It now has the opportunity to put policy into practice and devise a comprehensive disability scheme which will break the link between poverty and disability. It is vital therefore that it appreciates the groundswell of popular opinion that exists around revolutionising the welfare system. We ask therefore all our members to write to their MPs and also to Nicholas Scott and Tony Newton at the Department of Social Security. In your letters urge the Government to take note of the discrepancies that exist for people living with a disability aged dependent upon the welfare system.

At the same time ask them, whilst they are revising the system, to include people with disabilities and their representative agencies in the consultation process. When you hear back from your MPs the Society would appreciate receiving a copy of any correspondence.

This is a significant period for people with disabilities who are dependent upon Social Security for their income. It is therefore vital that we make the Government aware of our needs and enlist its support in enabling all people, regardless of disability, to have full access to every level of society.

Not Relevant

CAMPAIGNS

We **urgently** need people with haemophilia who are HIV positive, or who are widows or children of such people, to come forward for media interviews.

We will help you to understand some of the issues involved in doing this. Please contact the General Secretary, David Watters, as soon as possible.

We are turning away good media approaches every day because we need people to speak from personal experience.

EDITOR: Andy Cowe

EDITORIAL BOARD
Revd. A. Tanner MA
Andy Cowe

BLOOD TRANSFUSION SYMPOSIUM

14th International Symposium on Blood Transfusion, organised by the Red Cross Blood Bank Groningen-Drenthe, Groningen, NL, (three days) will be held in Groningen, The Netherlands on October 11-13, 1989.

The theme of the meeting is: **Cryopreservation and low temperature biology in blood transfusion,** under the chairmanship of Dr. Harold T. Meryman, ARC Rockville, MD, USA.

The meeting will have four sessions: I. Principles and fundamentals; II. Low temperature biology aspects; III. Cryopreservation aspects; IV. Advances and clinical applications.

Information and Registration: Symposium Secretariat, Red Cross Blood Bank Groningen-Drenthe, P.O. Box 1191, 9701 BD Groningen, The Netherlands. Tel/telefax: (0)50-137777. Telex 53942 AZGN.

Further details of the programme and speakers from The Haemophilia Society, 123 Westminster Bridge Road, London, SE1 7HR.

Plight of haemophilia in South West Africa

On Friday morning, 22nd October 1988, Prof Peter Hesseling, Tygerberg Hospital, Bellville, CP, travelled from Rundu in South West Africa (Namibia) to the Nyangana Catholic Mission Hospital on a dry, dusty, terribly corrugated road in a sizzling 38°C. He was supported by a grant from the SA Haemophilia Foundation and reported as follows:

The objective was to locate and examine GRO-A who had been the first Kavango child to be diagnosed as a classical haemophiliac in 1981.

He writes:

The kraal of the families lies about 2 km west of Nyangana Hospital and is perched on the southern bank of the picturesque Kavango River. I had a long interview with the family at the time of diagnosis, through an interpreter, and learnt that an older brother of GRO-A had died many years ago from uncontrollable haemorrhage after the horn of an ox had slightly grazed his cheek.

GRO-A showed symptoms of marked arthropathy. Severe bleeds had been treated with fresh blood by the missionary doctor at Nyangana Hospital, Dr. Barbara Potschka. Factor VIII concentrates and fresh frozen plasma were not available at the time.

Sadly, I learnt from his sister that GRO-A had been referred to Rundu Hospital in December 1984, following a severe joint bleed, and had died there on GRO-A GRO-A 1984. The exact cause of death had not been explained to them and I could not trace hospital records.

I was informed at Nyangana Hospital that another boy in the family had recently been diagnosed as a haemophiliac at Rundu Hospital. With GRO-A's sister, GRO-A as a guide, we visited her sister, Mrs. GRO-A GRO-A who lives 6 km to the west and is married to GRO-A GRO-A This couple have 4 boys named GRO-A GRO-A GRO-A Their three girls are named GRO-A

I mention their names to illustrate how complex names are in this society. GRO-A is the only haemophiliac amongst the four boys.

On our arrival his mother went to fetch him where he was playing 1 km away in the river. One could clearly see that his gait was abnormal as he approached from a distance. I estimated his age at 10 years. (The parents are illiterate and could not tell his age.) He has severe muscle wasting of both legs and arthropathy of both knee and elbow joints. Flexion of the right knee was limited to 70% and the left knee to 60%. Extension of both the right and left arms was limited to 10%.

The family (like most others in Kavango) are subsistence farmers and grow millet, have a few head of cattle, fish in the river and pick wild fruit in the forest. At present his only available treatment is still fresh blood. It is impractical, and for the family unacceptable, to send him to Windhoek, 860 km away, for long-term rehabilitation and education.

I have discussed his therapeutic requirements with the director of hospital services in Kavango and the haematologist in Windhoek, who agreed to order and make available dry Factor VIII at Nyangana and Rundu Hospitals for this patient.

I hope this brief report has illustrated the plight of haemophiliacs in remote corners of our sub-continent and will motivate members of the Haemophilia Foundation to investigate treatment facilities in other socio-economically deprived areas of our country.

I have obtained the names of 15 haemophiliacs in Namibia from my own records, records of the 3 paediatricians in the country and records of the haematology laboratory. I shall check these against the registered cases in the national register and will endeavour via the local social workers and doctors to arrange for better treatment facilities and care.

I gratefully acknowledge the support of the South African Haemophilia Foundation in this investigation.

(We are grateful to the South Africa Haemophilia Foundation for kind permission to reproduce this article from their May edition of 'The Haemophiliacs'.)

THE 1989 HAEMOPHILIA SOCIETY AWARD

Carol Holliday and Dr Tom Korn

As reported briefly in UPDATE No. 3, the 1989 Haemophilia Society Award was jointly presented to Mrs Carol Holliday and Dr Tom Korn who have both been the inspiration and the sustainers of the North Wales Adventure Holiday for young people with haemophilia. It was the Society's deep appreciation of those special holidays and the encouragement they have given to so many young people in living with haemophilia that led the Executive Committee to present the 1989 Award jointly.

In making the presentation at the 1989 AGM in London on Saturday May 27 the Chairman expressed his delight at being able to thank Carol and Dr Korn in this special way.

Carol's citation reads:

"Presented to Mrs Carol Holliday for her part in developing the North Wales Adventure Holiday in which she has been involved since its inception and for the careful attention she has given to the administration of this project so that the lives of boys with haemophilia have been greatly enriched."

Dr Korn's citation reads:

"Presented to Dr H E T Korn MB BS MRCPath for his initiative in establishing the North Wales Adventure Holiday and for continuing to give boys with haemophilia the opportunity to enjoy the freedom and sense of achievement this experience affords."

To both we offer our congratulations and thanks – and look forward to many more years of North Wales Adventure Holidays!!

RESEARCH ON TRANSGENIC ANIMALS

Many of you will have read and seen news items on the production of coagulation factor IX in the milk of sheep. This technique has been developed by John Clark and his colleagues at the Institute of Animal Physiology and Genetic Research in Edinburgh. It involves injecting the appropriate portion of human nucleic acid into a normal sheep's egg. These eggs are then returned to a surrogate mother and allowed to mature normally. A small proportion of the offspring incorporate the injected gene to yield a transgenic animal.

By injecting nucleic acid linked to an appropriate portion of animal nucleic acid, expression of the gene can be made tissue specific. John Clark's group have shown that by injecting the human factor IX gene linked to the gene for the major sheep milk protein beta-lactoglobulin it is possible to produce transgenic sheep that make small amounts of human factor IX in their milk, and that this characteristic is passed on to their offspring (see photo).

As in all scientific advances there are conceptual and development stages. The above studies have demonstrated that the concept of producing human protein in animals is valid and should be applicable to a range of therapeutic proteins in various animals. In fact, while farm animals have the attraction of size – they convert grass to large amounts of milk – such studies are costly and lengthy (sheep only produce lambs once a year).

Usually initial experiments are done in mice. Studies to date have demonstrated production of factor IX and one or two other therapeutic proteins in the milk of sheep and mice without any adverse effect on the animals. What is now required are the development stages – can the amount of protein in milk be increased to high levels and can it be purified completely from milk? A company, Pharmaceutical Proteins Ltd, has been set up to undertake these developments.

The chief advantage of this concept over the present methods for producing genetically engineered proteins (from cultured cells in the test tube) are that it has the potential for producing large amounts of recombinant protein from a renewable source at a relatively low cost – believe it or not it is a lot easier and cheaper to feed sheep than cells in the test tube.

C. V. Prowse

This picture, reproduced by kind permission of Scotsman Publications,shows some of the sheep involved in the experiments.

Not Relevant

WHAT DO YOU TELL A CHILD WITH AIDS?

GRO-D

reports from a meeting of experts on AIDS in children

The symposium on Paediatric AIDS, organised by the Haemophilia Society, was appropriately held in Scotland (Glasgow) where the experience of paediatric AIDS has been far more common than, as yet, in the rest of Britain.

The majority of paediatric cases so far have been among haemophilic children infected through contaminated blood products (factor VIII). The numbers of haemophilic children have now stabilised, the growing group being those children born to infected mothers.

Edinburgh, with its large population of HIV-infected drug users, has been the first city in the U.K. forced to come to grips with a growing number of infants born with HIV. Increasingly it will be this group of children that medical and social services will have to care for.

There are 320 reported cases of paediatric AIDS and HIV infection in the U.K. today. The real figure is somewhat larger. HIV tests for children, as Dr. Newell (Institute of Child Health) illustrated at the symposium, are still unreliable: it may be at least a year before a child's HIV status can be confirmed, but even then the progression of the disease is highly unpredictable.

Haemophilic children, by virtue of having appeared earlier and because their HIV status is easier to ascertain, are becoming increasingly well documented, and the progression of the disease better understood. It will be some time before there is a similar familiarity with HIV in newborns.

At the symposium were many of the most experienced doctors, social workers and health professionals in the field of paediatric AIDS, but even among the most "expert" of these there was confusion and uncertainty. Paediatricians wanted to know which was the better prophylactic, gammaglobulin or septrin? When should zidovudine treatment be introduced? Social workers and psychiatrists were asking: should you tell a child that he or she is HIV positive, and if so, when?

At these early stages of the disease among children in this country, questions such as these can only be answered, in the words of the Director of a day care centre for children with HIV in New York, on the basis that "this is one way of doing it, not necessarily the best".

POINTERS

New York will provide many pointers for us and may shed light on the development of paediatric AIDS in the U.K. There were an estimated 821 cases of HIV and AIDS among children in the city of New York in 1988, 91 per cent of those were born to black and hispanic mothers, and 80 per cent of children came from families where one or more of the parents were or had been intravenous drug users. We may be warned that paediatric AIDS could follow the same progression in this country and become a disease of the inner city minorities and drug users.

HIV is a disease of the family unit. For each infected infant there may be one or more parents of siblings dead, HIV positive or dying from AIDS. Assessment of the needs of a child with HIV must necessarily be family-centred. Some participants took a decidedly parent orientated approach to caring for children, and it came as a surprise to be refreshed by the child-centred approach of Dr. Mark Winter from the Isle of Thanet General Hospital in Margate.

Dr. Winter described his work in telling children of their HIV status; he described in particular the case of one infected boy who was intensely relieved to learn he had HIV; months of sitting outside waiting rooms while parents and doctors met behind closed doors had convinced the child that he had a disease so terrible he was afraid he would not wake up in the morning.

It became clear that caring for infected children will have to concern itself with drawing a careful balance between seeing the disease as a family problem and respecting the rights of the child.

A clash may be too strong a word to describe the difference between the medical and voluntary participants at the symposium over the use of the term 'high risk category'. It was a matter of some frustration to those voluntary representatives whose efforts have been precisely to escape from such terminology that the doctors continued to classify, and indeed act upon, the classifications of high risk or not high risk.

Dr. Jacqueline Mok, from the City Hospital, Edinburgh, presented a list of at-risk children that was so lengthy as to beg the question of who was *not* on the list, and if not why not? Isn't every child, like every adult, at risk if HIV infected?

Other points of language were also interesting, such as 'if a mother is particularly keen on breastfeeding her child I will *allow* her to do so'. Regret was expressed that many pregnant mothers were not found to be HIV positive until it was too late to have an abortion. It is regrettable that the mothers do not have a choice as to whether to continue with the pregnancy, not that termination wasn't possible. Non-discimination means respecting the desire of an HIV-positive woman to have a baby, regardless of how clinicians or anybody else may feel about that choice. HIV-positive pregnant women should be counselled much like any other mother at risk of passing on a hereditary disease, and their choice similarly respected.

NAT director, Margaret Jay, voiced the NAT's desire that voluntary childcare agencies should examine their potential roles as public educators, given their enormous range of contacts in the community. She spoke of the voluntary sector's abundant experience of providing high quality care for children with special needs. This expertise and experience will be called upon more and more as the numbers of children with HIV and AIDS grow.

RESOURCES

She expressed concern that policy institution and training was not as widespread as it might be. Our ideal, she said, was that "any member of any voluntary organisation that comes into contact with a child with HIV should be at least aware enough of the basic facts and issues to act in the best interests of the child". It is not enough, she added, to point out what should be done. Voluntary agencies should be empowered to address this issue with guidance and resources.

The Symposium was the first conference in the UK to address the issue of paediatric AIDS, and will be hopefully the first of more. While, as it became apparent, there is a need for dialogue across disciplines, a collective effort, within disciplines, to share information and knowledge was called for.

Not Relevant

NUCLEAR MEDICINE, HIV AND HAEMOPHILIA

Dr. M. O'Doherty

Department of Haematology, St. Thomas' Hospital

A year ago the Haemophilia Society awarded a grant to the Haemophilia Centre at St Thomas' Hospital. This grant was given to develop a non-invasive investigation to be used in the assessment of lung infections in patients with the Human Immunodeficiency Virus (HIV) and haemophilia. We wish to thank the Society for this grant and present some of the results we have obtained.

The most common infection associated with AIDS is an infection of the lungs called Pneumocystis carinii pneumonia (PCP), which develops in more than 60% of patients. This infection, unlike other pneumonias, may develop very slowly over a long period of time and in 5-10% of people may not be seen on a chest X-ray. To diagnose PCP, doctors have had to use investigations which until recently have involved looking into the lungs using a flexible fibreoptic telescope (bronchoscopy). This procedure has some risks and is not very pleasant. Recently the technique of obtaining sputum by inducing a cough has had encouraging results in making the diagnosis of PCP.

Having a bronchoscopy carries a low risk of haemorrhage in most categories of patients but, for obvious reasons, this risk may be high in patients with haemophilia. We have developed a technique in the Department of Nuclear Medicine to avoid procedures which carry the risk of haemorrhage. The specialty of Nuclear Medicine is often unknown to patients and the word NUCLEAR may have alarming connotations for some. Therefore before explaining our methods of investigation an outline of this speciality would seem appropriate.

WHAT IS NUCLEAR MEDICINE?

Nuclear Medicine is a branch of medicine involving the administration of small amounts of certain radioactive substances (radiopharmaceuticals) to evaluate medical problems. The radioactive material used is specially selected to be directed to the particular organ or part of the body to be studied i.e. lungs, heart, brain, kidneys, etc, allowing the specialist to assess the function of these parts. If the organ is not working properly then it is possible to demonstrate the extent to which particular parts are affected. The radiopharmaceuticals are safe, easy to use and can be administered by a number of different routes, either by inhalation, by mouth or by injection. The distribution and removal from the body of the radiopharmaceuticals can be observed using a gamma camera (as shown in picture). This camera "sees" the gamma rays (radiation) given off by the radiopharmaceutical and is linked to a computer system which provides detailed analysis of this information.

WHAT HAS OUR RESEARCH INVOLVED?

We have studied the removal of a particular radiopharmaceutical (99m Technetium Diethylenetriamine pentacetic acid) from the lungs of patients with HIV infection who have no symptoms as well as those with AIDS. This involved inhalation through a mouth piece of a small amount of radioactive substance as a fine mist for approximately 2 minutes whilst lying on their backs. The special couch (similar to the one provided by the Haemophilia Society) was then positioned over the gamma camera. Pictures of each patient's lungs were then recorded on a computer each minute for the following hour. After 40 minutes an injection of a small amount of radiopharmaceutical is given to allow various computer calculations to be made. The pictures that we obtained were different for smokers and nonsmokers. The smokers pictures are shown.

ST. THOMAS' HOSPITAL

The pictures show that the radiopharmaceutical inhaled into the lungs disappears from them over the next forty minutes (the lungs become more grey). This is not because the substance is breathed out but because there are small "holes" in the inside of the lungs, which allow the radiopharmaceutical to leak into the blood stream. The larger the holes, the faster the substances leak out. Thus for smokers who have bigger holes than nonsmokers half of the radiopharmaceutical leaks in 15 minutes whereas for nonsmokers this takes about 60 minutes.

GRO-A

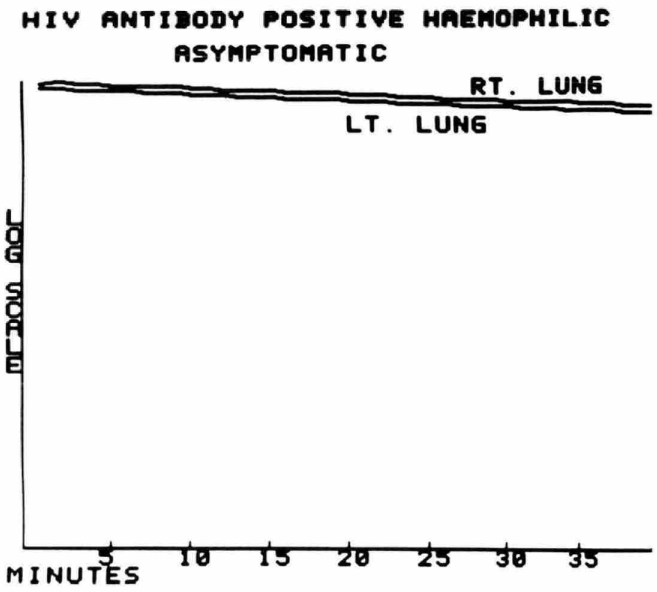
A patient undergoing treatment with the special apparatus.

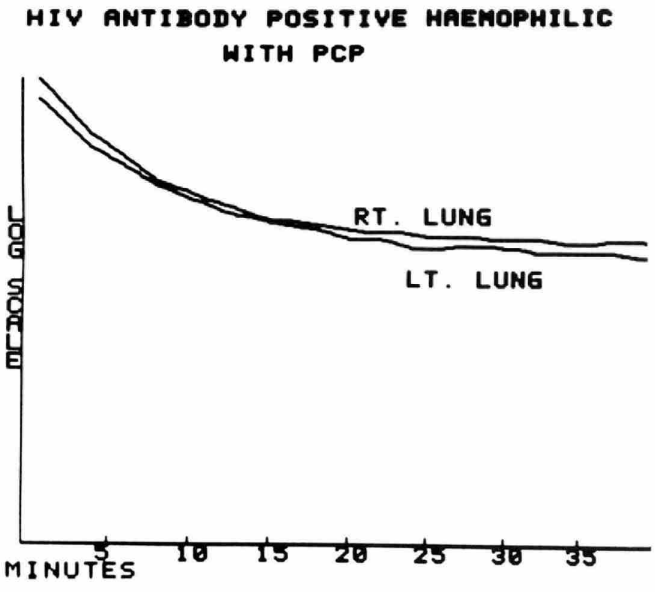
Using computer analysis, the times mentioned above are calculated and curves are produced showing the rate of disappearance of the radiopharmaceutical from the lungs. These are straight lines when drawn on a semilogarithmic plot in smokers and nonsmokers. We have found that the shape of these curves (lines) change in patients with Pneumocystis pneumonia (shown in picture) and the leak of radiopharmaceutical was very fast – taking approximately 2 minutes.

APPLICATION OF THE SCAN

The abnormal pattern and the very fast removal of the radiopharmaceutical was found in most patients with Pneumocystis pneumonia and not in other ordinary pneumonias. This finding is very useful since one can avoid the use of bronchoscopy in the early investigation of a patient who has a persistent cough or breathlessness and allow treatment to be started early. This test has been of particular benefit in making an early diagnosis of Pneumocystis, before abnormalities can be detected on a plain chest X-ray, and allowing either outpatient treatment or reassurance to be given appropriately. Should treatment be successful, then a repeat scan shows that the shape of the curves returns to normal (a straight line) after six weeks. This repeat investigation serves two purposes: firstly it confirms response to treatment but more importantly it also provides a scan for comparison if further problems occur in the future. This method has now been adopted into routine clinical practice in the hospital.

(Continued on following page)





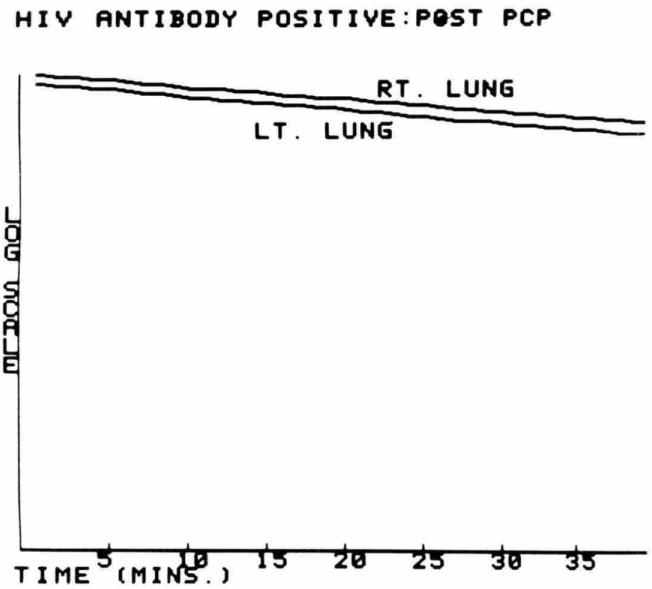
NUCLEAR MEDICINE

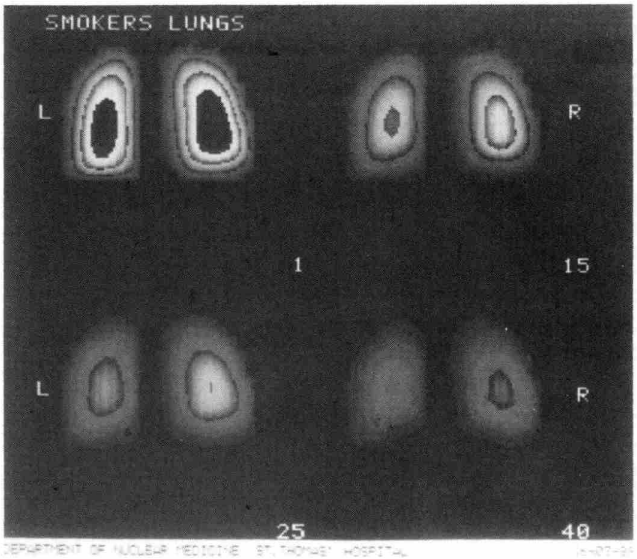
(Continued from previous page)

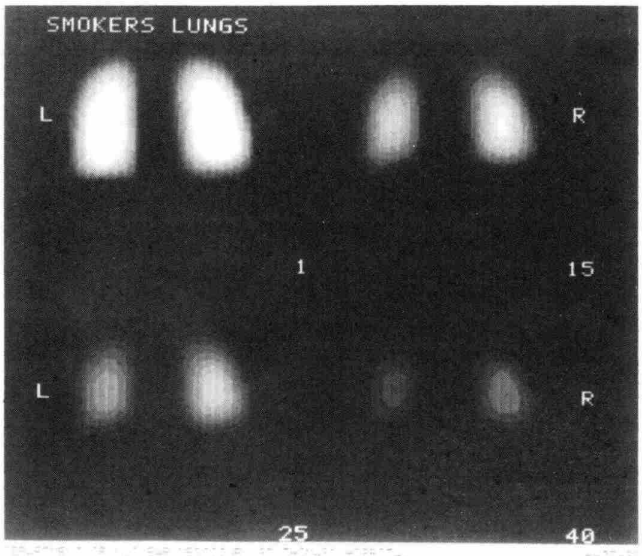
POTENTIAL USES OF NUCLEAR MEDICINE IN HIV INFECTION

The use of such small amounts of radioactivity is a safe and simple way of assessing how well different organs are working and has great potential in HIV disease to assess the function of the heart, brain, or kidney and the possible effects of drugs used in treatment on the function of these organs. The choice of particular radiopharmaceuticals to go to various organs will provide a means of assessing whether the antiviral agents are capable of altering the effects of the virus on these organs.

We wish to extend our particular thanks to all those patients who contributed to the development of this method of investigation and let them know that it is now in constant clinical use and has been of great value in the management of many patients with AIDS.







THE NORTHERN IRELAND HAEMOPHILIA CENTRE

ROYAL VICTORIA HOSPITAL, BELFAST

1958-1989

THE BEGINNING

The first step in the development of the Centre was taken in 1955 when a province-wide survey of patients suffering from hereditary disorders was undertaken. The results of the survey were published in 1958. It revealed that at that time there were 39 patients suffering from haemophilia A and only four suffering from haemophilia B. No other category of bleeding disorder was included. Subsequent to these findings, a special register was compiled and then facilities for diagnosis and management were offered to any such patients throughout Northern Ireland. Professor M G Nelson, then head of the Department, liaised with the Medical Research Council the body which was then responsible for haemophilia care in the United Kingdom and, under its auspices, the Royal Victoria Hospital laboratory became designated as the Haemophilia Centre for Northern Ireland.

THE PRESENT

In 1989 reappraisal of the register of bleeding disorders reveals a somewhat different picture. Although patients have died and others have emigrated, at present there are 125 patients registered with haemophilia A, 13 patients with haemophilia B, 11 patients with single inherited coagulopathies and 69 as suffering from varying degrees of severity of the von Willebrand syndrome. In addition, during the past two years, the register has been expanded to contain details of patients suffering from inherited platelet disorders. The increase in patient numbers has led to a gradual expansion of the number of personnel working in the Haemophilia Centre. The development of the Centre has paralleled the expansion of the Department of Haematology. The inpatient unit has 16 designated beds and for most, if not all, of the time at least 25% of these are occupied by haemophilic patients. In addition there is a Day Centre which operates Mondays through Fridays from early in the morning until 5 o'clock p.m. People with haemophilia are welcome at this Centre at all times but in particular there is a haemophilic clinic every Friday. A combined haemophilic/orthopaedic clinic is held on a Thursday afternoon every two to three months, depending upon demand. There is a non-emergency direct telephone line to the Centre for obtaining information, making appointments or leaving messages. At all other times a 24-hour on call service is operated from the ward.

Today it is difficult to realise that in the early 1950s there was no designated Haemophilia Centre, no Haemophilic Clinic, no special haemophilic register, no nurse, no physiotherapist, no dentist and no social worker whose main objective was the care of such patients. Therefore, a period of reflection can be beneficial as it enables one to appreciate the many changes which have occurred during the past 30 years. Gone are the days when people with haemophilia attended only on an emergency basis. In this Centre they used to come to the Haematology outpatient clinic which was held in the laboratory area within the Department of Clinical Pathology. The patients were admitted as and when necessary to any available vacant bed within the medical units of the Royal Victoria Hospital. Subsequently their care was shared between the ward physicians and the Haematology staff. There were no designated Haematology beds let alone any earmarked solely for haemophilic patients.

STAFF

The permanent medical staff of the Centre comprises three Consultant Haematologists: one, the author, who has overall responsibility for province – wide haemophilic care, aided by the Professor of Haematology, Professor J M Bridges, and Dr S I Dempsey, who carries responsibility for all paediatric activity in the Centre. Sister Catherine Farrell is the Sister-in-Charge of the outpatient facilities; she is on the committee of the Haemophilia Nurses' Association and is the present membership secretary of that organisation. She is ably supported by Staff Nurse Colette McAfee. In the ward, Sister Mary McGuigan keeps a close eye on haemophilia inpatient problems. Both are members of the Haemophilia Nurses' Association, as are many other ward nurses, making a total of seven members at the present time.

In 1983 a part-time haemophilic physiotherapist was appointed. Initially and for a further number of years she was funded by the Haemophilia Society. Mrs Lynne Crockard has proven her worth time and time again. Through her initiative and tenacity she has succeeded in persuading unwilling, severely affected patients to do their physiotherapy "tous les jours", rather than when "just in the mood". She is a founder member of the Haemophilia Chartered Physiotherapists' Group.

Mr David Kernohan has provided constant consultant dental care for many years. He runs a primary care clinic devoted solely to the management of patients with bleeding disorders. His attitude is firmly that "prevention" is better than "extraction". Other long-serving members of staff provide the essential laboratory back-up to the Centre. Senior Chief MLSO, John Carville, has been in charge of blood product provision ever since the late 1960s when the cryoprecipitate was prepared locally in the Department. After the transfer of its manufacture to the Northern Ireland Blood Transfusion Service, he became in some respects what might be described as "the keeper of the privy purse". He has never allowed the stocks of blood products to run dry and he and his staff in the hospital Blood Bank are responsible for issuing all the material for home treatment.

They maintain computerised records of the same and indeed all treatment – a job important and so necessary for the compilation of the annual returns for Oxford. No Haemophilia Centre can exist without accurate laboratory tests to establish the diagnosis and to maintain monitoring of the effectiveness of the treatment given and the Chief MLSO, Terry

NORTHERN IRELAND HAEMOPHILIA CENTRE (continued)

Ingles, has been in charge of the Coagulation Laboratory for more than 12 years. More recently new additions to the staff have taken place.

There is now a permanent secretary for the Haemophilia Centre, Mrs Colette Diamond. In 1987 a senior scientist, Dr Paul Winter, was appointed to help stimulate research and development. He is a molecular biologist and is engaged in expanding the diagnostic programme for carrier detection, etc. He is also interested in the molecular aspects of inhibitor patients.

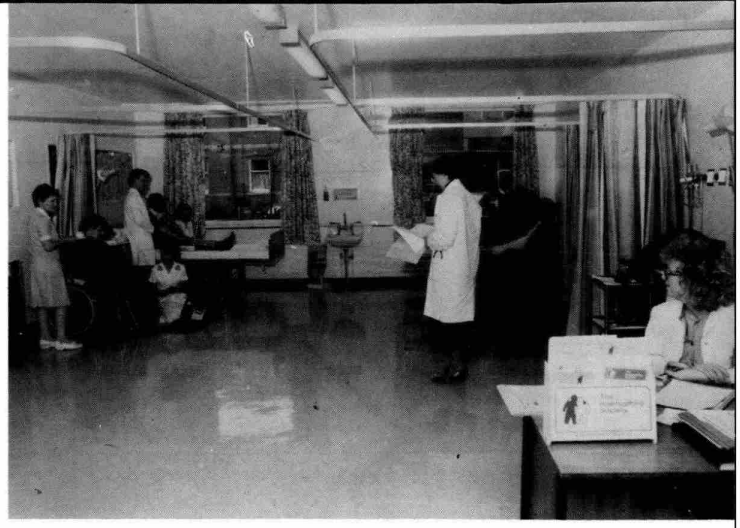
During the past five years the Centre has benefited greatly from the keen interest of R A B Mollan, Professor of Orthopaedic Surgery. He has enabled patients to have their orthopaedic surgery carried out locally and has contributed much through the combined haemophilic and orthopaedic clinic.

In the past the Northern Ireland Haemophilia Centre was not overwhelmed by help or interest in terms of input from social workers. Some came and went, some showed interest and occasionally when an excellent one was appointed, she had to leave, having gained promotion. Others declared that the problems of haemophilic patients were "insoluble". However, the picture during the past year has changed and since the appointment of Miss Geraldine Kerr, the social worker is now closely involved and is rapidly making her mark in the Centre and amongst the patients.

THE CENTRE

The Northern Ireland Haemophilia Centre serves a population of 1.5 million. Its patients are drawn from all over the Province and a reciprocal arrangement is operated between the Centres in Belfast and Dublin. This enables a small number of families from the Irish Republic to attend Belfast for convenience of travel, otherwise they would need to make a long and tedious journey to Dublin. In return, patients from the Northern Ireland Centre who go on holiday to the Republic of Ireland are treated free of charge at appropriate southern Centres. Since the inauguration of the Centre in 1958, it is easy to highlight the outstanding changes in the patterns and methods of haemophilic treatment. Perhaps it would be logical to list them chronologically but as most readers are familiar with the sequence and saga of events, it may be more pertinent to comment briefly on the activities of the Centre under the headings of **Initial happenings, DIY, prophylaxis, orthopaedics and the future.**

Initially the Centre's aims were directed towards the provision of an efficient 24-hour service for treating acute bleeding episodes. Once this was firmly established under the control of personnel well versed in the management of the haemophilic patient, the direction of the Centre's policy was altered towards the DIY approach. Thus the home care programme flourished from the mid-70s until the early 80s by which time all the patients who were suitably motivated, and had attained Sister Farrell's required level of aseptic expertise, had been recruited. Of course, each year one or two new patients join the club and there are now 50 patients on home treatment.



'Headmistress reading the lesson', or The Friday Haemophilia Clinic.

After the home care programme was firmly established, the theme of prophylaxis was introduced and, although prophylactic factor VIII injections were included in the programme, as for example, to break a cycle of repeated joint bleeds, they did not constitute its main objective. This was directed more towards prophylactic physiotherapy, dental hygiene, regular follow-up and a general educational approach towards a more active life-style. At some time during the 1970s the Centre adopted a motto, namely: "Happiness is haemostasis". It sounds banal and, of course, cannot be applied to traumatic bleeding but, over the ensuing years, the patients seemed to achieve benefit from its implied philosophy. In practice, it meant that non-therapeutic injections of factor VIII or IX were taken (or given if not on home treatment) before times of known or possible stress, e.g. one schoolboy always bled just before his family was due to go on holiday and frequently caused delays and general family upset. Other examples are well-known, i.e. before school or other exams, on the morning of an interview or on the night before the wedding. All Centres practice such "insurance" treatment but the patients in this Centre seem keen to cling both to their insurance treatments and to their motto. At the present time much of the major activity in the Centre has been directed towards the management of surgical problems, both general and orthopaedic in nature. Professor Mollan plans two to three major orthopaedic operations each year and carries out other more minor procedures as and when necessary. The general surgery cases over the past few years have included a total colectomy, parathyroidectomy, prostatectomy, cholecystectomy and repair of hernia, etc. It is hoped that this spate of surgical activity will not continue, as it is extremely expensive in terms of factor replacement therapy.

THE FUTURE

Patients remain worried regarding the complications of the treatment of their inherited bleeding disorders. They have been largely reassured about the efficacy of heat treatment, pasturisation and other techniques used to diminish the viral contamination. However, they still remain worried about their future, therefore much of the Centre's activity is now directed towards giving "time" for the patients. It takes a long time to discuss all the topics, whether related to HIV infection, hepatitis, the safety of the treatment, future treatments, if with monoclonal factor VIII, genetically engineered factor VIII, the problems of the inhibitors and their treatment, etc. Therefore, it seems the most important treatment at present is to give time for discussion of problems and, at least in comparison to the early 1950s, the 1990s seem to offer a better prospect to the patients.

AUTHOR: E E Mayne, MD FRCP FRCPath, Director, Northern Ireland Haemophilia Centre.

GRO-A

Patient and mother's first visit to clinic for diagnosis of mild bleeding disorder and staff nurse Colette McAfee and Dr. Helen Magennis, senior registrar.

Not Relevant

Not Relevant

Not Relevant

Not Relevant

BOOK REVIEW

Understanding Haemophilia

A Personal Account and Practical Guide for Parents, Teachers and the Caring Professions

– GRO-A –

Published by
Ashgrove Press
Price £4.95

In publishing this book, GRO-A GRO-A has, in her own words, provided the sort of information she would have liked to have at her fingertips when her son was first diagnosed.

The book begins with the story of Mrs GRO-A s experiences when her son, GRO-A was diagnosed as having haemophilia. She explains her feelings and the family's reactions in the first few months, and then goes on to explain how, as time went on, the whole family adjusted to life with haemophilia.

Mrs. GRO-A s book is full of sound practical advice and reliable information interspersed with personal experiences. It is interesting and entertaining and gives an encouraging, hopeful and happy view of living with haemophilia.

This book is highly recommended, in conjunction with information available from the Haemophilia Society, as an excellent source of reference for anyone interested in haemophilia or involved in the care of a child with haemophilia.

Ashgrove Distribution,
4 Brassmill Centre,
Brassmill Lane,
Bath BA1 3JN.

HAVE YOU EVER WONDERED WHAT HAPPENS TO YOUR BLOOD?

This article is reprinted from 'National Haemophilia' the journal of the Haemophilia Foundation of Australia, who, in turn, reprinted the article with the permission of the Red Cross Blood Bank, Victoria.

Blood is made up of red cells, white cells and platelets suspended in a clear yellowish fluid called plasma. Contained in the plasma are vital substances essential for good health e.g. sugars and proteins.

WHOLE BLOOD TRANSFUSIONS

Whole blood is used in in transfusions for the replacement of blood loss during surgery and for the treatment of severe haemorrhage in accidents and maternity cases. In such cases the red cells in the transfused blood improve the oxygen supply to the tissues and the plasma restores the volume of blood to the amount the body needs to maintain normal blood pressure.

USE OF BLOOD COMPONENTS

There are many conditions which do not always require the transfusion of whole blood. By separating whole blood into its components and using the particular blood component needed by a patient it is now possible to use a single donation to treat several patients suffering from different illnesses. This technique is known as blood component therapy.

When the Red Cross Blood Bank laboratory team has completed the initial task of separating the main blood components, the remaining plasma is further subdivided into its constituent parts, the so-called plasm fractions. The process of plasma fractionation is carried out at the Commonwealth Serum Laboratories.

The preparation of blood components is based on a simple principle – the fresher the blood the better the results and the more efficient use of each donation. To achieve the best results the interval between the collection of blood from the donor and the commencement of laboratory processes should be as short as possible. It is for this reason that we have a special courier available to collect blood from our Mobile Units in suburban areas and transport it back to the Central Bank at various times during the donor session. In this way we can process many extra donations for blood components and concentrates. Similarly, when donors attend the Bank in the morning (8 a.m.) we are able to commence the necessary procedures for blood component therapy at an early hour so that the components are ready for distribution the same day if necessary.

The following is a list of the blood products available from a single donation:

Fresh Frozen Plasma. This is the fluid portion of the blood, separated and frozen immediately after collection. It is used to treat patients who have a generalized clotting deficiency.

Platelets. Platelets are produced in the bone marrow by the breaking up of large cells called megakaryocytes. They act as plugs to seal off small holes in blood vessels and also take part in the clotting process. They are used for the control of haemorrhage in patients whose platelets have become defective or deficient.

Cryoprecipitate. This product is prepared from fresh frozen plasma and it contains a blood clotting substance called Factor VIII, which is absent in patients who suffer from haemophilia.

White Cells. Fresh concentrates of white blood cells are sometimes given to patients who are not producing their own, or they may be used for the production of interferon. Some scientists think that interferon could be a cure for some forms of cancer, multiple sclerosis or even the common cold.

Red Cell Concentrates. These are prepared by removing part of the liquid plasma from each donation: the remaining concentrated red cells are ideal for the treatment of anaemia if transfusion is required.

Frozen Red Cells. Freezing is used for the long term storage of rare types of blood and also for the removal of all traces of white cells from blood which is required for patients who are allergic to white cells.

Filtered Red Cells. Special filters may also be used to remove white cells from blood for patients who have antibodies against them. The white cells cling to fibres of cottonwool or cellulose acetate as the blood passes through the filter, but the red cells pass through unaffected.

By Fractionation From The Plasma –

Normal Immunoglobulin. An extract from the plasma which carries the antibodies against common infectious diseases. Used to give temporary protection against diseases such as measles, rubella and infectious hepatitis.

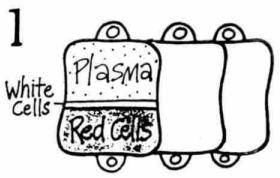
Hyper-Immune Globulins. These are prepared from a pool of donations from donors who have strong antibodies against a particular disease. They are used for such things as the treatment of tetanus or the prevention of conditions likle hepatitis B, herpes zoster or haemolytic disease of the newborn.

Albumin. A solution of the main protein present in human plasma. Used in the correction of protein deficiency especially that associated with kidney and liver diseases, toxaemia of pregnancy and open-heart surgery.

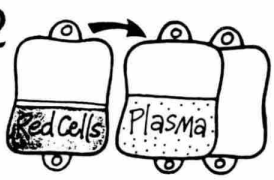
Stable Plasma Protein Solution (S.P.P.S.). This is a solution mainly of albumin but it also contains some of the globulins of the plasma. It is used for the treatment of shock, often while waiting for compatible blood to be obtained. As it does not have any living component, it can be stored for up to five years and most hospitals in all parts of the country have reserves of S.P.P.S. for emergency use.

Anti-Haemophilic Factor (A.H.F.). Like cryoprecipitate, this preparation contains the blood clotting substance called Factor VIII in a dried, concentrated form. The availability of this product has revolutionized the management of haemophilia in recent years.

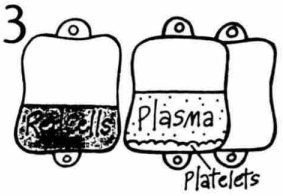
Prothrombinex. A concentrate, rich in coagulation Factors II, IX and X extracted from plasma by a chemical process and subsequently freeze-dried. Used for treatment of haemophilia 'B', otherwise known as Christmas Disease, and other conditions where there is a deficiency in any of these factors.



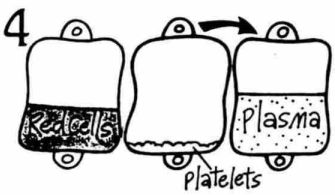
1
The blood donation is collected into a plastic pack which has two or three satellite packs connected by plastic tubing. The blood in the main pack is placed in a centrifuge and spun at 1800 revolutions per minute for seven minutes so that the red and white cells gravitate to the bottom of the pack.



2
The plasma, which contains the platelets and clotting factors, is now at the top of the pack. By squeezing the pack the plasma is forced into one of the satellite packs.



3
The pack is then spun again in the centrifuge, at a higher speed so that the platelets are concentrated at the bottom of the pack.



4
The plasma is then squeezed into another satellite pack where it is ready for further sub-division into its constituent parts – e.g. anti-haemophilic factor, fibrinogen, immunoglobulins, albumin, stable plasma protein solution.

ZIDOVUDINE OR AZT

Guide for patient use

Adapted from an article produced by Harborview Medical Centre AIDS Clinic.

WHAT IS ZIDOVUDINE?

Zidovudine (RETROVIR, and known as AZT) is an antiviral drug made by a company called Burroughs Wellcome. It is used to treat people who have disease caused by Human Immunodeficiency Virus (HIV). It has been shown to prolong the lives of persons with AIDS and many studies are exploring the effects of zidovudine in other HIV-infected persons. Zidovudine is not a cure.

HOW DOES ZIDOVUDINE WORK?

To understand how zidovudine works requires knowing about microbiology and physiology. To explain all the details is beyond the scope of this guide. We will give a simple explanation of its action.

One of the white blood cells that HIV invades is the T4 lymphocyte. T4 lymphocytes normally help the human body fight infections by "organizing" the immune system when an infection occurs. HIV prevents the T4 lymphocyte from organizing other cells of the immune system and may eventually kill infected T4 cells. HIV uses T4 lymphocytes to make more HIVs. HIV can spread to other T4 lymphocytes and decrease the total number of T4 lymphocytes that a person has. This process of infection and destruction of cells may repeat itself again and again until eventually there are very few T4 lymphocytes left. Then the body cannot fight very well against certain kinds of infections or cancers. This condition is what is know as the Acquired Immune Deficiency Syndrome or AIDS. Other cells besides T4 lymphocytes can also be infected by HIV.

It is important to realize that zidovudine will not kill HIV and will not repair damage already done to T4 lymphocytes or the immune system. It is not, therefore, a cure for AIDS. It can slow down destruction of cells and may permit other immune building processes to occur. It is an effective treatment.

HOW IS ZIDOVUDINE TAKEN?

Zidovudine is a white capsule with a dark blue band. It is made in one strength only (100mg) except for research purposes. The usual starting dose is 200mg (2 of the 100mg capsules) every four hours.

WHAT KIND OF SIDE EFFECTS OCCUR WITH ZIDOVUDINE?

The most common side effects are:

1. Bone marrow problems
2. Nausea
3. Headache
4. Muscle pain
5. Difficulty sleeping

1. **Bone Marrow Problems**

The bone marrow is responsible for supplying the body with: Red Blood Cells (RBC's) that carry oxygen to the body; White Blood Cells (WBC's) that protect the body against infection.

Zidovudine may suppress the function of your bone marrow. If your RBC level drops, you become anaemic and may need a blood transfusion to build your RBC level to normal. If anaemia is severe, you may get very tired. There is not any easy way to transfuse WBC's, so if you get a low WBC on zidovudine, you may need to decrease your dose, or temporarily stop taking zidovudine until your WBC returns to higher levels. When someone is taking zidovudine, blood counts should be done regularly to monitor these counts. *It is important to keep all of your clinic appointments so that any adverse effects can be detected and treated early.*

2. **Nausea**

Many of the people taking zidovudine experience nausea at first. Most people say that it goes away or they get used to it after a few weeks. Others say it comes and goes as long as they take zidovudine. Many people with AIDS or ARC have already lost weight and cannot afford to lose any more. To help decrease nausea and to prevent loss of appetite try these suggestions:

a. Take your zidovudine with some food, such as dry biscuits.
b. Before getting out of bed in the morning, eat a few biscuits, a handful of dry cereal, or a piece of toast or dry bread. Keep these within reach of your bed.
c. Get up slowly in the morning. Avoid sudden movements.
d. Eat smaller, more frequent meals throughout the day to avoid an empty stomach, which can make nausea worse.
e. Eat slowly.
f. Drink fluids (including soups) between, rather than with, meals.
g. Avoid strong-smelling foods. Sometimes cold foods are more appealing because they have less odour.
h. Avoid your favourite foods when you feel nauseated, so that you do not later associate these with feeling sick.

3. **Headache**

For mild headaches, Panadol is often helpful. These headaches often go away after several weeks. For more severe "pounding" headaches, talk with your doctor. These sometimes occur when people are anaemic.

4. **Muscle Aches**

If you experience muscle pain or aching you may take Panadol. If the pain persists, talk with your doctor.

5. **Trouble Sleeping**

If you have trouble sleeping after the first few weeks of taking zidovudine, you can try the following:

Omit alcohol and beverages with caffeine, especially in the afternoon and evening; try a cup of warm milk or herb tea before bed.

Take a warm bath before bed.

Slow down before bed by getting a back rub, listening to soothing music, or reading something boring.

If you continue to have problems sleeping, discuss with your doctor the possibility of taking a sleeping medication.

HOW DOES ZIDOVUDINE MIX WITH OTHER DRUGS?

Zidovudine is metabolized or "burned up" in the liver and "washed out" of the body by the kidneys (in the urine). Many drugs are handled in this manner and may compete with each other for elimination if they are taken chronically. When there is competition between drugs, higher than normal levels of drugs may build up in the body and cause problems.

Please check with your doctor before taking other drugs.

HOW MUCH DOES ZIDOVUDINE COST?

Estimated average retail price for the standard dose of 200mg (2x100mg capsules every four hours equalling 360 capsules per month) is about $10,000 per year. (£5,000 approx).

HOW DOES ZIDOVUDINE AFFECT LIFESTYLE?

Taking zidovudine means that you care about your health. Lifestyle may have a large impact on your health and you may want to explore the following issues:

1. Recreational drug use – Do you use or abuse recreational drugs? Do you know why you use drugs? Is it for relaxation. physical pleasure, or to escape? Are there healthy things you could substitute for drug use?
2. Diet – What foods do you enjoy? Do you have a problem gaining or maintaining weight? Is your diet well-balanced and does it include all four food groups?
3. Activity – Are you too active or not active enough? Do you allow yourself enough rest between activities?
4. Mental – Do you take care of your emotions? Do you spend time with people who make you feel liked and loved? Do you have healthy ways of coping with stress in your life?
5. Spiritual – What is sacred to you? Do your beliefs comfort you or do they cause conflicts within you? What gives you meaning and hope in your life? Where do you turn in times of despair?

Resources to help you explore these issues include family, friends, co-workers, nutritionists, massage therapists, clergy, health care professionals and others.

(Acknowledgements to National Haemophilia, The Journel of the Haemophilia Foundation of Australia).

Not Relevant

From the Paleolithic Period

A member recalls his experience

This year I shall be seventy and a reasonably accomplished adaptee to haemophilia. I read with interest in the May issue of THE BULLETIN the account of experiences twenty years ago. You think things have changed?

I can tell you about events sixty-five years ago in the Paleolithic period of treatment.

We had the usual "classical" family history of my mother's brother etc. etc., but what on earth can be classical about such a deviance I have never appreciated. Events were shrouded in mystery; the topic had an enormous taboo placed upon it as though some kind of incestuous in-breeding had produced this enormity. Victoria, the transmission-belt to the crowned heads of Europe, with her nine children and thirty-four grandchildren, was the key to the attitude. Close intermarriage was the cause . . . or so ran the popular adage.

I once researched the memoirs of diplomats to the Russian Court at the time of the revolution and found their guarded comments about haemophilia and the role of Rasputin to be hilarious. According to them I should have been dead years ago; I was lucky. My Mum had no Rasputin around. Often the condition was not mentioned by name but as "a serious affliction". Maurice Paleologue in his memoirs commented: "It is supposed to be a sign of degeneracy" . . . forgive my smiles.

At one-and-a-half (and before my conscious memory) I had my first experience of hospitals when I cut the thin ridge of tissue between the upper lip and the gum (the Frenum), rich in blood vessels and a vulnerable area for the haemophilic child crawling around with toys in the mouth. The treatment was apparently repeated stitching and pressure. With gratification I do not remember it.

'Ignorance of internal bleeding'

The astonishing thing is that there was total ignorance of internal bleeding as a symptom and the fear was always of external haemorrhage. It was the middle of the nineteen-twenties before I had prolonged spells in hospitals. The first one was when I had a milk tooth pulled out by a dentist because it was causing problems for its successor. This was the precursor of treatment for the next thirty years; repeated stitching, pressure on the gum and my jaws bandaged-up with strong rubber ropes to maintain the pressure.

Such barbarities are not to be recommended for the growing child.

The stitching was repeated whenever the clots broke down and bleeding began again in earnest. The spell in hospital lasted three weeks but parents were not allowed to visit! The regulations stated one parental visit per month because it was assumed traumas for the child resulted. James Robertson (whose research started with work on children evacuated during the war) overturned all this rubbishy thinking by revealing that the opposite was true. Modern concepts recognise the long-term pyschological damage which can be done to children separated from their mothers at times of stress. We now have beds provided in hospitals for mothers to be near their children.

My boyhood had periods of parental separation which were profoundly painful experiences. Medical doctors had little knowledge of the haemophilic condition and a sense of ignorance is very quickly transferred to the patient – even if he is very young. It is then that panic takes over. On such occasions I knew that doctors had not the foggiest idea what to do with me.

This is not a question of profound insight; even a six-year old had a simple understanding of what they were saying to each other as they talked across me when I was lying in bed. When I was discharged for the first time after the bleeding from a tooth socket, the doctor in charge at Leeds General Infirmary solemnly told my mother that I had "spongy gums". When my mother, with working-class commonsense, drew attention to the inherited tendency she was told that the gum condition was an inherited one!

The usual falls of boyhood with massive haematomas, and the pain which acts as an accompaniment, proved far too great a problem for the family doctor. He prescribed hot poultices to prevent sepsis! Today we pack it with ice and in retrospect I wonder how we survived with such counter-productive treatment. I had an extended spell in hospital in the nineteen-twenties after breaking a thigh.

The conventional treatment included traction for a couple of months and every time a nurse walked down the ward it was standard practice to pull the ropes upon which the weights were suspended over the pulley at the end of the bed. With a thigh which was a mass of congealed blood, and a trigger-point for the kind of pain which only haemophiliacs can describe, I could not disguise childish yells. For this I was derided as a "softy" but the pain was nothing compared with that of separation from my parents. They were warm, loving and intelligent. Every day in hospital they wrote to me in simple words which a boy could understand. They kept me in the land of the living; the hospital system could so easily have crushed me.

When finally discharged, my father bought a second-hand pushchair and fitted a wooden support for my leg. I wore a caliper splint for the best part of a year. This had a ring round my groin and was intended to take the weight off my leg. It had the good effect of preventing further bleeds, but the leg is still vulnerable and I have had it encased in plaster three times since the event. If T. S. Elliot could say that "My life is measured out in coffee spoons", I feel I have an equal right to say that mine is measured out in hospitals I have known. The problem afterwards was one of making my leg bend again . . . a task to which my father applied himself nightly with more enthusiasm than understanding. I still remember the regular encounters with the Inquisition and the thought brings me out in a cold sweat. However crude it may have been, my father succeeded; I can bend my leg, I have no limp and there was no physiotherapy department to advise or help him.

It was the beginning of the Schools' health service and supervision and my leg and dropped arches were spotted by the school doctor who came to our Elementary School in Holbeck. Professor Richard Hoggart has with great insight described the sociology of the area in *The Uses of Literacy.* Three mornings a week I visited the School Clinic for massage by a nurse. This middle-aged, dewy-eyed lady was a member of the Mazdaznan religious sect who believed in forms of strength-through-joy and participation by exercises and singing. She massaged my feet for twenty minutes on each visit and sang "Up The Airy Mountains, Down The Rocky Glen" as an accompaniment. The erotic effect on a growing boy was profound, even if he was a haemophiliac.

I still, however, have flat feet.

'I could not disguise childish yells'

There were no peripatetic teachers in hospitals before the war. Children were left to rot, in educational terms. My parents made up for all that by encouraging me to read and write under their tuition and when eleven I got a scholarship to the local Grammar School. This was so unusual an occurrence that my school gave everyone a holiday . . . the only benefit I ever conferred upon them. My mother and father gave me this chance by devoting so much time to me, when they also had two younger girls to look after. The mother of a haemophilic boy always carries an enormous burden. His pain tugs at her heartstrings and at the same time she feels guilt at having transmitted the gene which gives her no personal problems. She endures all the pain of her boy and feels responsible for it. The Czarina can be forgiven for Rasputin who, by hypnotism, seems to have been able to help her son.

On later occasions my mother would take me back to hospital for the umpteenth time after a tooth extraction and the nurses would go through their routine of changing the bedclothes after they were blood-soaked. Matrons were far keener on outward cleanliness than assuaging the haemorrhage of a frightened boy. Mum would sit at the bedside making plugs for me to bite upon, consoling me with tears in her eyes and holding my hand to give me courage and comfort. In due course a doctor would appear to stitch me . . . yet again, and she would be banished back home. My record was sixteen times in one socket but my mother's anguish could never be measured.

At Grammar School I was excused rugby . . . but not boxing,

(Continued on next page)

which gave me a bad bleed from my left ear and a legacy of deafness. By the time I was eleven the mysterious name haemophilia was recognised and I had become some kind of a freak. The term is not exaggerated, for scholarship boys who did not conform were not given the easiest of rides. Teeth, internal bleeds from bumps (a whacking one from a cricket ball) all conspired to make me different. Boys do not want to be different.

They want to be accepted by their peers. I spent several periods in the Infirmary and fell behind at school. I dropped-out. My Mum and Dad could do little to help me. They were self-educated and knew little of Algebra and Latin. My father got me an apprenticeship as a printer and I left school at fourteen and a half. He had to pay £10 penalty to the Education Committee; this was more than three weeks' wages. There are those today who pay to send their children to allegedly good schools. My father paid to remove me from one!

'It scared me to death'

It was then, in the late twenties, that I had my first blood-transfusion. It was person-to-person from a fireman and scared me to death as a strange kind of umbilical cord carried his blood in to merge with mine. It all ended with an almighty rigor and I can but think that the blood-grouping had not been correctly sorted out. This experience underlined my oddity and I felt condemned to a lifetime of deviance. The worst feature of pre-war medical conditions was that no-one ever tried to provide explanations. The patient was not entitled to knowledge; he should be gratified by the rudimentary treatment. Blood tests were unknown to all the hospitals I frequented.

Apprentice life was tough for we walked to the factory, worked for 8 hours, and then hared off to the "local Tech." for evening classes, but as one grows older haemophilic fears recede. Boys become streetwise and learn to avoid bumps. One learns the vulnerable places and protects by reflex action the linkage joints. I launched into evening classes with great enthusiasm and discovered that by working I could more than hold my own. This coincided with my first taste (an apt description) in Leeds Infirmary of Russell's Viper Venom. After a tooth extraction it was painted onto sockets and layers of gauze were impregnated with this first real clotting agent. The taste was vilely bitter and the mouth soon filled with lumps of acrid liver. Doctors regarded it as a miracle break-through; patients with some resignation accepted that at last someone seemed to be trying to help them . . . or poison them. I had so many varied treatments in the fifties and sixties that I have lost count of them. Oxycell, reputedly extracted from Bulls' testes, had a vogue at one time, but was about as effective as the other bullish extract.

The part-time route to education is a test of stamina more than ability. I collected sufficient technical qualifications to enable me to move into colleges as a teacher, but the habit dies hard. I have noticed with other haemophiliacs that the sense of inner resentment is a great spur to action. I managed a first degree and then a (taught) Masters, along with other qualifications which helped me to keep pace with technological change. I cannot remember any period in my life when I was not attending an evening class in something or other. My wife and I have just returned from Budapest, where I gave a talk on the British system of Further Education, to Hungarian teachers. They were intrigued by the multiplicity of "do-it-yourself" facilities available for late developers. We thought ourselves trail-blazers as apprentices and our formal evening classes were supplemented by Workers' Educational Association and National Council of Labour College Courses, sponsored by our trade union. I notice that today the NGA assists its members with Open University Fees and continues the tradition of working class self-education.

'Even cryo didn't solve my problem'

Lecturing in a college had one great advantage. I could use the long vac. for medical treatment and until Factor VIII came along I usually needed most of the holiday if I had a tooth extracted. Even Cryo did not solve my problems, though it helped a lot. I'm probably the only man to go into hospital to have two teeth extracted, who came out six weeks later on crutches with my leg in plaster after a bleed from an intramuscular penicillin injection. Put that in *The Guinness Book of Records!* For twenty-five years I had a nagging hernia. If ever I was in hospital and, after a long battle, some doctor managed to restore me to normality, he invariably said he would "like to have a go at that hernia of yours". I notice they said little about it when still baffled by my bleeds.

Being a born-again coward, I always declined with (I hope) politeness until Factor VIII came on the scene and the op. became a necessity. 'Twas a miracle. In two weeks I was back at work . . . but with hepatitis after it. This was my first experience of the Elixir and I thought that all our problems were finished with. Little did we know ... A Biochemist colleague warned me in the early days about possible dangers and for six years I refused treatment. This was the best decision I ever made and was possible only because I knew I could survive without it. My boyhood told me so. I have however run the whole gamut of treatment from nothing to (hopefully) safe Factor VIII . . . it is not really a claim to boast about.

My grandchildren look upon me as the original Bionic Man. I have a steel plate through my left shoe so that I can walk with a minimum of pain from a stiffened joint. I have a hearing aid for use in important committee meetings and I have a splint for my right hand to ease the problem of driving. That is a legacy of a handshake from a masculine 'hearty' and I have had so many internal bleeds into it that I now either proffer the left hand or brush the finger-tips with regal elegance. Impressive male handshakes are a curse. As Zsa Zsa Gabor said "Men who are macho are not often mucho". The Spanish royal family padded the tree trunks in the garden where a haemophilic son could play; I pad my left thigh. We adjust at differing levels.

Despite the assaults upon the National Health Service, our European services for haemophiliacs are supreme. When working in a Caribbean island, advising a Minister on the reorganisation of a college, I had a bad bleed into a foot when I turned sharply in a hole in the road. In a flash I was back to my boyhood but with a little more experience. Having seen the state of technology I was determined not to venture near to a hospital; I had three weeks of pain and gloom before I managed to win the tussle with the aid of the fridge. I knew then that I was alive because I lived in Europe. The Third World must kill off most of its haemophiliacs just as surely as Hitler tried to rid himself of the biologically undesirable.

The strange thing is that sixty-five years later I have less trouble with haemophilia than ever I did as a boy. In many ways, the challenges it set proved to be a positive advantage, but I still regret the lost glories of youth. I wanted so much to play football, but above all I wanted to be normal. There is one possible consolation. Possibly the only person to have a City and Guilds Silver Medal for a craft examination and a Ph.D. will prove to be a person with haemophilia.

Not Relevant

YOUR CHILD IN SCHOOL

by Carol Brown

Choosing the right school

You have the right to choose which school you would like your child to attend. Start thinking early and approach your Local Education Authority (LEA) for details of schools in your area. Every LEA has to publish general information for its area.

Contact schools you are interested in and arrange to meet with the Headteacher and look around the school. Talk to parents.

Take special note of the physical layout of the school building. Are there a lot of stairs and how might this affect your child if his mobility is restricted as a result of a bleed?

When you have chosen

In most cases a child will be offered a place in the chosen school. Occasionally, a particular school will have more applications than places. Should your child be turned down for any reason, you have the right to appeal. The appeal is heard by a special committee and, if it decides against you, you should be informed of the reasons. No school should exclude a child on account of his haemophilia!

What to tell the school

Do inform the school clearly about your son's haemophilia. Be positive, and let them see that most of the time your child has exactly the same needs as his peers, but occasionally he may require specialist attention for his condition. The main role of the school will be to contact the appropriate person as these needs arise.

Special Guidelines for schools have been drawn up by the Haemophilia Society, to be issued to every LEA. You can obtain a copy from the Society to give to the headteacher of your child's school. These Guidelines give a clear explanation of the needs of a haemophilic boy and how the school should respond to these needs.

If your son is HIV positive

The decision about whether to tell the school is entirely yours. You are under no obligation to do so, and you should think very carefully before divulging this information. If the school requests your son's HIV status, refer them to the Guidelines, which state quite clearly that this information is not relevant.

Problems in school

Most boys have very few problems in school. Any minor troubles can usually be sorted out between parents and school. However, there may be certain situations where a boy has his mobility permanently or frequently restricted due to bleeding episodes. This may result in frequent absences, or an inability to move around the school to gain access to certain classrooms. First you should approach the school to see if they can help out. In the case of short-term absence, work could be sent home.

Where the difficulty is more long-term, or the school is unable to provide resources (such as ramps) necessary for your child, you may wish to consider having your child's needs assessed towards the making of a **statement of need.** The school may suggest this procedure, but there can be drawbacks. The 1981 Education Act introduced this procedure to ensure all children with "special needs" receive education appropriate to their needs.

A panel of professional advisers is drawn up by the LEA to consider the assessment. You have the right to attend any examination and to be informed and consulted at all stages. LEA's are required to take account of any representations made by parents. You will be given a copy of the proposed statement, and if you are unhappy with any part of it, you can request a further meeting of the panel to discuss your worries.

If the LEA decides to go ahead with the statement after the meeting, you can appeal to the Secretary of State for Education and Science. You may wish to do this if, for instance, the LEA decides that the needs of your child cannot be met at the school of your choice and he must go to a school or special unit they consider appropriate.

You must abide by the decision of the Secretary of State. However, the vast majority of haemophilic children should never need to be statemented, and the procedure should not be considered without careful consultation between parents, school and Centre staff.

The school and you

It will help your child enormously if you take an active role in the life of the school. If there is a PTA, do join. Schools always need willing parents to help out in certain activities – perhaps in the school library, or on school outings. You may even wish to consider becoming a parent-governor, where you can have considerable influence on the running of the school.

Support and encourage your child by taking a keen interest in his work. Attend open days and talk to your son's teachers about his progress. Continue to take an interest throughout his school career.

Changes in schools

Many changes are taking place in our schools at present as a result of the Education Reform Bill. There is no reason why these changes should have an adverse effect on the education of the boy with haemophilia. Good education is largely due, as it has always been, to a co-operative partnership between home and school, working together to enable the child to achieve to his full potential.

The changes resulting from the Reform Bill are outlined in a handbook "Our Changing Schools, A handbook for parents". All parents of children in school should have received a copy, but if you would like one, they can be obtained, free of charge, from DES, Publications Despatch Centre, Government Buildings, Honeypot Lane, Stanmore, Middlesex HA7 1AZ.

For further information concerning your rights in schools contact:—

Advisory Centre for Education (ACE), 18, Victoria Park Square, London E2 9PB. 01-980 4596.

Not Relevant

DRUG TREATMENT TRIALS AT DIFFERENT STAGES OF HIV INFECTION

This is a brief and incomplete listing of drugs at different stages of testing for patients with HIV infection in the USA.

Research studies are categorized by the extent of research (Phase I, II and III), defined as follows:—

Phase I is the first stage of human testing in small numbers of people at very few locations. Phase I studies are designed to establish a safe dose range for the drug, to gather information on the action of the drug inside the body and how long a dose of the drug acts on the body.

Phase II trials are studies that evaluate the effectiveness of the drug or combination of drugs and usually include several hundred people. These trials are designed to assess the effectiveness of the drug in a precise manner, as well as to detect major adverse reactions.

Phase III testing can involve large numbers of volunteers. These studies must confirm earlier findings of effectiveness and should detect side effects that occur only rarely or after a long period of treatment.

FDA Food and Drug Administration

IND Investigational New Drug

AL-721
Status: Investigational: Phase I/II studies in progress
Mechanism: extracts cholesterol from invading virus cell membranes rendering them unable to penetrate host cells and reproduce
Side effects: slight diarrhoea and nausea

Aerosolized Pentamidine
Status: Approved as treatment IND
Mechansim: an antiparasite drug, useful in *Pneumocystis carinii* pneumonia
Side effects: cough/bronchospasm, fatigue, and burning sensation in the back of the throat

Ampligen
Status: Investigational: Phase I/II studies in progress
Mechanism: antiviral: stimulates immune system and suppresses growth of HIV by increasing interferon production
Side effects: mild flu-like symptoms

AZT/Retrovir
Status: FDA approved for AIDS: Phase III studies in progress for asymptomatic persons
Mechanism: antiviral, inhibits reverse transcriptase, an enzyme which is critical to the reproduction of the virus
Side effects: bone marrow suppression, nausea, myalgia, insomnia, headaches, anaemia, leukopenia, neutropenia, abnormal liver function

Amphotericin B
Status: Phase I studies in progress
Mechanism: antifungal antibiotic, binds to sterols in viral membranes, thereby affecting membrane permeability and causing loss of viral infectivity
Side effects: headache, anorexia, weight loss, nausea, vomiting, diarrhoea, muscle and joint pains and reversible renal dysfunction

Acyclovir
Status: FDA approved drug: Phase I/II/III studies with AZT in progress
Mechanism: antiviral effective in preventing recurrent herpes infections
Side effects: nausea, vomiting, headaches, diarrhoea, dizziness, anorexia, fatigue

Alphap-Interferon
Status: FDA approved: Phase I/II/III studies in progress
Mechanism: antiviral protein that has been found effective in treating patients with Kaposi's sarcoma. Believed to be capable of reducing reverse transcriptase and interface with a final stage of viral reproduction
Side effects: fever, fatigue, chills and headaches

Colony Stimulating Factor-G CSF
Status: Investigational
Mechanism: shown to be a powerful stimulator of mature human neutrophils:
Side effects: mild fever, myalgia, headache, nausea, skin rash and diarrhoea

Dextran Sulfate
Status: Investigational: Phase I/II studies – (Phase II will be in progress shortly)
Mechanism: anticoagulant which theoretically inhibits reverse transcriptase and interferes with the attachment of HIV to T4 cells
Side effects: lowers platelet count, loss of appetite and mild diarrhoea

DTC (Imuthial)
Status: Investigational: Phase I/II studies in progress
Mechanism: stimulates the liver to produce a thymic hormone-like activity called hepatosin. Theoretically this helps speed maturation of T4 cells, enhance overall T cell functions, improves T4:T8 ratio and slows the reproduction of HIV
Side effects: stomach cramps and nausea.
Precaution: DTC cannot be used when alcohol has been consumed.

Isoprinosine
Status: Investigational: Phase III studies in progress
Mechanism: synthetic immunopotentiating agent, enhances macrophage activation, natural killer cell activity and production of lymphotoxin *in vitro*
Side effects: no side effects reported

Peptide T
Status: Investigational: Phase I studies in progress
Mechanism: attaches to the receptor site of the surface of the T cell preventing the virus from attaching
Side effects: no side effects reported

Ribavirin
Status: Approved by FDA for its use in aerosol form for the treatment of respiratory syncytial viral infection. Double-blind placebo controlled trials in progress
Mechanism: is a synthetic nucleoside
Side effects: mild anaemia, elevated bilirubin, insomnia, headache and irritability

IMREG-I and II
Status: Investigational: Phase II/III studies in progress
Mechanism: immune regulator: Imreg appears to regulate the immune system by stimulating interleukin-2, and gamma interferon:
Side effects: no side effects have been reported

Foscarnet
Status: Investigational: Phase I studies in progress
Mechanism: has *in vitro* antiviral activity against all human herpes viruses: mode of action is selective inhibition of viral DNA polymerases
Side effects: headaches, nausea and anorexia

Dideoxycytidine (DDC)
Status: Investigational
Mechanism: inhibits HIV reverse transcriptase activity
Side effects: development of peripheral neuropathy in some HIV infected patients

Recombinant CD4
Status: Investigational
Mechanism: has been shown to inhibit HIV infectivity, replication and virus induced cell fusion, *in vitro:* is believed to achieve such inhibition by acting as a soluble virus receptor which interferes with the binding of HIV to intact CD4 molecules on the surface of target cells
Side effects: no side effects reported to date

Acknowledgements to
Hemophilia Information Exchange: AIDS Update,
24 April 1989. National Hemophilia Foundation,
New York, USA

This item is published for the interest of our readers and does not imply endorsement by The Society nor does it indicate that the drugs are all available in the UK.

1992: HAEMOPHILIA AND EUROPE

The UK Haemophilia Society is currently co-ordinating the work of The European Haemophilia Consortium. This is a new group of all the European Haemophilia Societies which met, for the first time, in Noordwyke in the Netherlands earlier in the year.

The Consortium's first major task was to stop some particularly dangerous proposals being put to the European Parliament. These could have totally disrupted European supplies of blood products after December 1991. Much vigorous activity was put into this by our own Jonathan Cooper and GRO-D from the Netherlands (known to many of us as the Netherlands representative at WFH meetings).

So excellent was their work on our behalf that all the proposals were scrapped and we hope that the European Consortium will be consulted in future about such proposals. Our activities were also recognised in an article in Time International on 'Eurolobbying'.

Our photos show some of the delegates at the conference in the Netherlands.

Not Relevant