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The Bulletin

Professor Bloom speaks in Amsterdam

IMPROVEMENT IN PURITY ESSENTIAL

At the XIIIth Congress of the International Society of Thrombosis and Haemostasis held in Amsterdam, Professor Arthur Bloom, director of the Cardiff Haemophilia Centre, had several points to make that were of interest to people with haemophilia.

Speaking at the Congress, he said that improving the purity of factor VIII or factor IX therapy is a priority, especially in those with HIV infection. It has been claimed that less pure concentrates down-regulate the immune system in a manner unrelated to HIV itself. This is thought to result from the action of unknown protein impurities. Although clinical evidence is sparse, it has been found that HIV positive patients who are switched to purer treatment show a slowing down in the gradual reduction of their T4 lymphocyte count. Professor Bloom urged switching to purer concentrates, noting that this would, however, have economic implications.

MAINSTAY OF TREATMENT

Recent production of purified factor IX containing less contaminants may be expected to produce improvements in use but this has not yet been proven in practice. "However, there is no doubt that it should become the mainstay of treatment of haemophilia B," said Professor Bloom.

Professor Bloom also pointed out that recombinant factor VIII is undergoing trials by two companies and that it has been shown to be effective in preventing bleeding.

DEVELOPMENT OF INHIBITORS

He noted that the development of inhibitors in six previously untreated cases had been worrying, but that the inhibitors have not generally prevented continuation of treatment. It remains to be seen whether this will be a significant problem.

Professor Bloom also said that considerable experience is accumulating with recombinant factor VIIIa which acts independently of the intrinsic system by bypassing factors VIII and IX to activate factor X. He said that the results had been reasonably successful and that factor VIIIa seemed to be quite promising.

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In this issue

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In this letter from America GRO-A *describes . . .*

THE LIFE OF A PERSON WITH HAEMOPHILIA IN THE USA

Greetings to all UK people with haemophilia. My name is GRO-A **I am from the United States and have haemophilia. I have been asked to write a short article for your Bulletin describing the life of a person with haemophilia in the US.**

This life is, I am sure, in many ways similar to the life you lead in the UK. We have many of the same concerns in the US as you do in the UK. The haemophilic community in the US has been rocked by the AIDS virus, just as yours has. In this way I am certain that we are very similar. However, due to the great differences we have in our systems of health care, the way we address these concerns is markedly different. These differences will become more apparent as you read this article.

Before I begin to describe life for people with haemophilia in the US, I would like to tell you about myself. I am a 24 year old with classic, moderate haemophilia who is HIV positive. I live in GRO-A Michigan. I have lived my whole life in the state of Michigan. In 1990 I received a Bachelor of Science degree in Nursing from the University of Michigan. In July of that same year I married my wife GRO-A Since getting my degree I have worked in the Coronary Intensive Care Unit of the University of Michigan Hospitals as a Registered Nurse.

There are 20,000 people with haemophilia in the US. This is a very small segment of the American population. Of that 20,000, 17,000 have haemophilia A, 2,800 have haemophilia B, and 200 have other assorted clotting factor deficiencies. An estimated 70 per cent of the people with classic haemophilias are severe, 15 per cent moderate and 15 per cent mild. Of those with Christmas disease, an estimated 50 per cent are severe, 30 per cent are moderate, and 20 per cent mild. People with severe haemophilia in my country, both A and B, use 60,000-80,000 units of factor annually. The moderates use 15,000-25,000 units annually and the milds use 2,000-5,000 units annually. At least two thirds of people with haemophilia in the US treat themselves at home. The rest, many of them children under 10, are treated by their centres or emergency rooms.

'We suffer a great deal of joint deformities'

As you can tell from the amount of factor we use in the US, we are not fortunate enough to be consistently treated on a prophylactic basis. Our capitalistic form of health care does not allow us to have the same luxuries as our German and Swedish brethren. The people with haemophilia in the US, thus, suffer from a great deal of joint deformities. I myself cannot straighten either of my elbows fully. My ankles creak when I walk and my left knee has lost some of its mobility.

'No exact numbers as to the cases of hepatitis'

In this way we are very similar, even if our forms of health care differ.

The most sobering numbers facing the US haemophilic community are those associated with the HIV virus. The National Haemophilia Foundation (NHF), our equivalent to the UK Haemophilia Society, estimates that 50 per cent of people with haemophilia currently suffer from HIV infection. Many in the community feel that this estimate is low. They feel that there are more like 80 per cent of all people with haemophilia infected. Whether or not the NHF numbers are low, the figure is still staggering. It has, however, begun to plateau and even drop in this country. This is due to the unfortunate deaths of 5 per cent of the HIV infected people with haemophilia, the use of AIDS drugs, and the widespread use of monoclonal factor VIII in this country.

The percentage of HIV infected people with haemophilia dying from aids is lower than other US populations affected by the disease. This is probably due to the availability of health care services to people with haemophilia, the quick and appropriate use of AIDS drugs, and the advent of pure monoclonal products. I am happy to report that there are some people with haemophilia born after 1987 who have used nothing but monoclonal factor VIII in their lifetimes.

Hepatitis has long been a nemesis of the haemophilic community in the US. Although new cases of the disease have been all but wiped out by the advent of the monoclonals, there are still some new cases of the disease coming from the segment of our community who are being forced by financial or discriminatory circumstances to use intermediate purity products. There are no exact numbers as to the cases of hepatitis in my country, but they are very low because most of the haemophilic population is currently using monoclonals.

People with haemophilia in my country have a national, private, non-profit organisation which works for the interests of people with haemophilia and Von Willebrand's nationwide. This organisation is the National Haemophilia Foundation (NHF). The NHF is further split into state foundations. These

GRO-A

GRO-A

state foundations work for the interests of the people with haemophilia and Von Wildebrand's who reside in that state.

The NHF is organised like any other corporation in the states. It provides many services to the people of this country with haemophilia on a national basis and is involved heavily in lobbying at a national level. New outreach and support groups such as Women's Outreach Network (WONN) and Mens Advocacy Network (MANN) have been organised by the NHF. These groups are intended to empower people with haemophilia and their significant others with the ability to start support groups in their region or state.

The NHF also has the Haemophilia AIDS/HIV Network for Dissemination of Information (HANDI) which is an arm of the organisation devoted to disseminating information.

With all the work that the NHF does on a national level it is unable to reach each person with haemophilia in this country on an individual level. This is where the state foundations come in.

The state foundations are independent organisations which are not directly tied to the NHF. They do, however, carry out the work of the NHF at a state level. They also have their own programmes and projects which are unique to their state or region. The state foundations lobby to their state governments regarding state issues which will affect the lives of people with haemophilia. Their link to the NHF is loose, at best. They pay the NHF for the services it provides them at the national level. Finally, the NHF sets general guidelines by which the state foundations must operate to stay within the organisation. As you can see, even though they are separate entities the NHF and the state foundations are linked together by both commitment to the haemophilia community and by loose financial and idea sharing ties.

The individuals who work closest to each person with haemophilia in the US work in the treatment centres. There is at least one treatment centre in each state. They are funded by federal grants which are administered by the state foundations to the treatment centre. Thus there is also a loose association between the state foundations and the treatment centres.

The clinics are comprehensive in nature. When a person with haemophilia attends one, depending on the size and funding of the clinic, he will be seen by his haematologist, nurse co-ordinator, social worker, dentist, physical therapist, and possibly a health care professional who works directly with HIV and related issues.

Research has been done regarding the use of monoclonals and its effect on CD4 counts. A meta-analysis of these studies proves that the use of monoclonally produced factor by people with haemophilia has either prevented a rapid decrease in CD4 counts or has actually caused an increase in these counts.

I, myself, began using monoclonally produced factor in 1987. At the time my CD4 counts were around 200. One year after beginning to use monoclonally produced factor my CD4 count had risen to 578. This is without the use of AZT or any other adjunct medication. It has remained greater than 500 since that time. I firmly believe that monoclonally produced factor VIII should be used to treat EVERY person with haemophilia in the world. To treat a person with classic haemophilia with anything less is outrageous.

The practice of treating certain people with haemophilia with less than monoclonal factor does, however, occur in the US. Some physicians and government agencies have deemed the use of monoclonal factor VIII unnecessary for people with haemophilia who are HIV positive. They treat these people with haemophilia, mostly those who are poor and must depend on government aid for their health costs, with heat treated or detergent washed products.

This is done mainly to reduce the cost because monoclonals are more expensive than the intermediate purity products. The physicians who do this believe that there is no use wasting monoclonals or the money needed to buy them on a population with a fatal disease.

This is called discrimination in my country. However, those who practice this form of discrimination are never challenged. They are powerful people who are drawn to a higher, more noble cause, saving the government money.

Life for a person with haemophilia in the US is affected by many of the same things that affect your life. The only major differences between our lives and yours is the product of differing government and health care systems. Our blood, our hurdles to overcome, and our grief at the thought of fallen brethren is probably much the same. I wish you well as you fight to overcome your hurdles. Stay healthy and never stop fighting.

A letter from America

'The product of differing government and health care systems'

GRO-A will be in the United Kingdom from November 18 for a few days to meet with local groups.

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PRODUCT PURITY

The question of high purity factor concentrates is one that is of immediate concern to everyone with haemophilia, here 'The Bulletin' reviews the current situation.

Recent research findings have established that clotting factor concentrate infusions, in and of themselves, suppress the immune system because of the presence of foreign proteins. The purer the product, the less the suppression. Product purity, aside from the issue of having HIV-free factor, is important to the person with haemophilia because of the impact that each infusion has on the immune system, whether that individual is HIV-positive or negative.

EXPERT OPINION

In this report we look at product purity and give the opinions of three American haemophilia experts.

We begin with Dr Peter Levine MD who is the former director of the Comprehensive Haemophilia Treatment Center in Worcester, Massachusetts and has served as medical director of the National Haemophilia Foundation. He gives his views on blood product purity and the immune system.

"During the first years of the AIDS epidemic, in the period between approximately 1983 and 1985, we and other investigators began to gather data on the nature of the immune abnormality in people with haemophilia," said Dr Levine. "In a series of studies we were able to demonstrate that the immune abnormality in people with haemophilia was multi-factoral. Part of the immune abnormality was due to the HIV virus. Another part was independently attributable to certain other viral illnesses such as hepatitis virus in persons with haemophilia. Interestingly, however, there was also an immune abnormality attributable to the intensity of exposure to intermediate purity factor concentrates. This strongly suggested that there was something else about the factor concentrates capable of producing abnormalities of lymphocyte function, and leading to altered immunity.

"It is important to remember that 'intermediate purity' concentrates are, in fact, not terribly concentrated. They contain less than 1% clotting factor concentrate and more than 99% extraneous and unnecessary proteins from the many donors who have contributed to the pool. On the other hand, the new high purity products, after purification by monoclonal antibody technology, are less than 1% impurities and more than 99% pure clotting factor, prior to the addition of albumin. Albumin itself is a pasteurised protein with a remarkable record of safety and no known effect on the immune system.

HYPOTHESIS TESTED

"In order to test the hypothesis of whether high purity clotting factor concentrates would be better for the immune system we have carefully followed the immune studies of a group of seven patients who were given high purity clotting factor concentrate, and they continue to do somewhat better than comparison groups receiving intermediate purity concentrate.

"Although a variety of viral kill technologies have been introduced in the intermediate purity products the fact remains that large quantities of foreign proteins remain in these materials. These foreign proteins include the killed viruses. This is quite different from the pasteurised high purity products, where the foreign proteins and killed viruses are removed subsequent to pasteurisation.

"All of this gives rise to the interesting hypothesis that the high purity products may in fact be the treatment of choice in individuals whose immune systems are already abnormal on the basis of human immunodeficiency virus infection. It will be several years before the definitive answers are in.

COMMON SENSE

"In the meantime an increasing number of treaters are moving towards the more purified products, largely on the basis of the common sense approach that it is hard to believe that purer is not better."

On the subject of selecting a concentrate we turn to David Green MD PhD who serves as the director of the Northwestern Haemophilia Center in Chicago.

"Performance is determined by how well the product works. The potency of a concentrate is listed on the label; through trial and error, you learn the dose of clotting activity that works best in controlling your bleeding. You can then select the product that has a potency that most fits your needs.

BOTTOM LINE

"On the safety question the bottom line when considering concentrates is that the product is not contaminated with infection-causing viruses.

"Today's concentrates are treated in a variety of ways to kill these viruses. These include heating and/or adding detergents; these methods destroy the HIV virus and probably most of the hepatitis viruses. Also, some concentrates undergo several additional purification steps which raises the question of whether these high purity concentrates are safer than the old intermediate purity concentrates.

"High purity concentrates are more expensive than intermediate purity concentrates. If a concentrate works well for its user, and causes no deterioration in his immune system, I would see no reason to make a change. I would recommend that an individual continue using the product that has proven effective in the control of his bleeding episodes, but that he has regular checks on the level of his immune cells. If these begin to deteriorate despite the use of AZT or other anti-viral agents, then a switch to a high-purity concentrate should be considered."

INTERMEDIATE PURITY

On the question of when should an intermediate purity factor be used we turn to Maria Gordon MD who is the associate director of the Haemophilia Comprehensive Care Centre at the Children's Hospital of Los Angeles in California.

"There is sufficient evidence available today that high purity factor VIII concentrates do not affect the immune system adversely. Improvements of a haemophilic person's immune function can occur within six months from discontinuing use of impure factor concentrates and simultaneous treatment of bleeding episodes with high-purity products.

"Haemophilia treaters commonly prescribe these new high-purity factor concentrates for previously untreated patients or those who show no evidence of HIV or hepatitis virus infection. In my opinion the safest and most effective therapy should be made available to all patients regardless of HIV status or presence of hepatitis virus-induced liver disease."

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THE WORLD FEDERATION OF HEMOPHILIA

What it means to us in the UK

Declan Murphy

"The Federation has benefited immensely from the splendid medical expertise of doctors, nurses and medical personnel from the UK"

At the recent Annual General Meeting of the U.K. Haemophilia Society, guest speaker Declan Murphy, the Executive Director of the World Federation of Hemophilia, covered three main topics.

These were: the importance of the UK Haemophilia Society within the World Federation of Hemophilia, the function of the Federation, and the philosophy of the Federation.

On the importance of the UK Haemophilia Society within the Federation he said, "As you all know, it was one of the original founding members in Copenhagen in 1963, alongside five other countries. We are also fortunate, like you, to have Alan Tanner as our Chairman. He offers to the Federation, credibility, stability and a marvellous link with the tradition of the Federation.

Over the years, the Federation has benefited immensely from the splendid medical expertise of doctors, nurses and medical personnel from the U.K. The organisational ability of the U.K. Haemophilia Society has deeply influenced the structure, constitution and procedures of the World Federation. And for all of this, I would like to express in the name of the Federation and its members our deepest gratitude."

On the work of the World Federation he said, "I am happy to say it continues to expand. Three new members were admitted to the Federation in Washington:

The Federation continues to 'further care and treatment'

Czechoslovakia, the People's Republic of China and the USSR — they are all members who are beginning to organise themselves and certainly need the example and support of established national members such as you.

"The Federation continues to further the care and treatment of people with haemophilia through its 22 International Haemophilia Training Centres. Last year, nine fellowships were given to medical personnel from seven countries.

"In recent years, the Federation's relationship with the World Health Organisation has grown. Two members of the Federation are on the global Blood Safety Initiative Programme. Last year, a joint WFH/WHO Faculty, under the leadership of Prof. Mannucci wrote a paper entitled 'The Possible Control and Prevention of Haemophilia in the Developing World'. The Federation has also collaborated with the Global programme on AIDS by writing two state-of-the-art texts on haemophilia and HIV, and is now preparing a pilot programme on haemophilia and AIDS to be held in Santiago, Chile in September.

"Hopefully, we can have subsequent workshops in other developing countries to help people confront the reality of haemophilia and AIDS. The Federation has also been invited by WHO to participate with a panel discussion on haemophilia and AIDS at the VIIth International AIDS Conference in Florence in June."

On the philosophy of the Federation he said, "It is, in one word, a philosophy of service. In a recent study of state-of-the-art haematologic treatment, speaking of the objectives of treatment, it says: 'The objectives are to minimise disability and prolong life, to facilitate general social and physical well-being, and to help each patient to realise their full potential'.

"The roll of the Federation is to stimulate:

1) The medical world to offer the best and most comprehensive treatment to people with haemophilia.

2) To stimulate industry to offer a safe and affordable product to people with haemophilia.

3) To stimulate people with haemophilia to be fully themselves, realising their full potential, never feeling different or less.

"Yes, the eighties were terrible and terrifying years. Yes, they have left us a horrible heritage. Nonetheless, the only life we know, and the only life we have, is today. The today that I live and breathe and that life today is our challenge. Bitterness and resentment serve no purpose. They make us less. It is up to each of us to reach into the marvellous resources of our spirit to confront life and live it to the fullest.

'Bitterness and resentment serve no purpose — they make us less'

"I would like to quote from a recent book by a young Irish man with haemophilia and AIDS. His name is [redacted] and he says: 'The answer is belief in my own strength and power. With every battle I fight I get stronger. I have come to terms with the past. And I live in the present.'

"The Italians have a marvellous saying: 'What is important is not to add years to your life but life to your years.' In the end, what makes a difference in life is not how much we have or what has been allotted to us in life. It is our attitude.

"My final invitation to you is that this new world political atmosphere of freedom, where there are so many wonderful opportunities to make a difference, in this extraordinary new political unity of Europe where the world is getting smaller, we need to work together. I would like to invite you to consider the World Federation as a vehicle, a significant vehicle for greater involvement in the world community."

A look at last year

This year's Annual General Meeting of The Haemophilia Society was held at the Nevin Theatre at St Thomas' Hospital.

At the meeting the Chairman, The Revd. Preb. Alan Tanner, said, "One of the points to note from the past year was the conclusion of the HIV litigation. While acceptance lay with the lawyers, and the lawyers only, the outcome would not have been possible without the support of so many people for the Society's campaign.

"Haemophilia and Safer Sex" was published during the year and won first prize in the Domestos Health Education Awards. The prize money has been earmarked for future educational publications.

"1991 also saw the introduction of Haemofact HIV Treatment News, which has been warmly received. This is an occasional publication which will be produced as the need arises.

"In March the Society held its first weekend for women affected by HIV through partners, children, etc., in Newcastle. A similar conference was held in Durham in April this year. The Member Services Committee are exploring new formats for conferences for the families of those affected by HIV directly or indirectly.

NHS REVIEW

"Through the Policy Committee we have been looking closely at the impact of the NHS Review on the treatment and care of haemophilia. We fear that the outcome might not be promising and it is important that we monitor and make public our justifiable concerns about the future.

PRIORITY AREAS

"The Society's priority areas of activity in the future will include looking at hepatitis and the ways in which it will affect people with haemophilia. Other priority areas include the NHS Review; the safety and purity of blood products and, perhaps most important of all, the building up of our local Groups and the overall improvement in relations between the national office and our local representatives."

The Haemophilia Society Award 1991 was presented to Sister Maureen Fearns of the Newcastle Centre. She has done a great deal for people with haemophilia in the UK over the past 20 years and has worked on the development of care for patients.

The Haemophilia Society Award for outstanding service, to acknowledge the vast contribution made to the work of the Society within our local Groups was presented to [redacted]. She has been an enthusiastic member of the Council for many years and has worked hard to ensure that her local Group continued in the face of many difficulties.

A Haemophilia Society award was presented to the General Secretary to mark the sterling work he did throughout the HIV compensation litigation.

The R G Macfarlane award will be presented to Professor Arthur Bloom on Friday October 25 at the Cardiff Centre.

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HAEMOPHILIA AND HEPATITIS PAST, PRESENT AND FUTURE

Christine A. Lee, MA, MD, MRC, Path, FRCP

Presented as the keynote address at the Annual General Meeting.

"HEPATITIS" means inflammation of the liver and for people suffering from haemophilia this is most commonly caused by past viral infections.

PAST: The first description of the clinical features of epidemic jaundice were made by Hippocrates who lived from 460-475 BC. He recommended a special diet of "melikraton" which is a mixture of water and honey as a treatment.

An outbreak of jaundice, which occurred amongst workers in a Bremen shipyard in 1883-4, demonstrated that hepatitis could be transmitted by blood. It was thought that this was due to the vaccination using human lymph because 191 out of 1289 vaccinated who fell ill had all been vaccinated against smallpox on the same day in August 1883.

USING CLINICAL OBSERVATION

In 1943, using clinical observation, Beeson described seven people who had developed jaundice following blood transfusion. He suggested that the illnesses were due to the transfusion of the blood. Amongst his recommendations were that a careful record should be kept of the source of plasma or blood and that "a small portion of blood or plasma should be set aside at the time a transfusion is given so that in the event of subsequent cases of hepatitis some of the causative material will be available for study."

During the Second World War the blood transfusion service in this country became firmly established. It also enabled a study of the relative risks of single donor and multiple donor blood products. An analysis was made of records of blood products issued from the North West London area from 1940. Pooled plasma from pools varying in size from 30-200 litres had been used in the treatment of burns and crush injuries sustained during the bombing of London. It was found that human plasma for treatment should not be pooled. This work was published in the British Medical Journal by Spurling and colleagues in 1945.

PROGRESS MADE

Important progress was made in 1963 when a description of the first viral marker of hepatitis was made. This was the "Australian antigen" described by Blumberg. He showed that the blood from a multi-transfused person with haemophilia (who must have had a circulating virus of Hepatitis B): hence the name "Australian antigen".

The Willowbrook School experiments distinguished infectious hepatitis (hepatitis A) from serum hepatitis (hepatitis B).

More recently it has been shown that the hepatitis that 50,000 US servicemen developed in World War II was caused by hepatitis B. Most of the veterans tested in 1985 showed antibodies to hepatitis B. It was thought that this outbreak could be linked to a specific lot of yellow-fever vaccine.

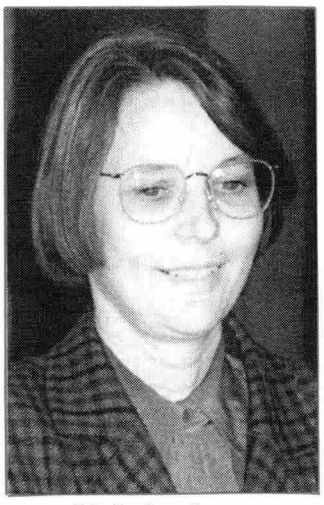
During the 1960's cryoprecipitate, which was rich in factor VIII, was discovered by Judith Pool. Gradually, patients with haemophilia A were able to have home treatment with this product. Each bag came from a single donor so that the risk of hepatitis was low, especially since all blood donations were tested for hepatitis B.

POST-TRANSFUSION HEPATITIS

However, during the 1970's lyophilised or freeze dried factor VIII and IX were made. These came from donations of 10,000-20,000 donors. There was thus a 100% incidence of post-transfusion hepatitis in people with haemophilia who were treated with these products for the first time. This was so-called non-A non-B hepatitis because it was neither hepatitis A nor hepatitis B. The virus has been identified only in the last two years as hepatitis C (HCV). From 1985 both factor VIII and IX have been sterilised by various methods — mostly heat —and there is now no longer risk of transmission of hepatitis or HIV by these blood products.

PRESENT: At least five viruses have now been identified which cause hepatitis: A, B, C, D, and E. However only B, C, and D can be transmitted by blood products.

All donor units of blood are now tested for hepatitis B. Most older people with haemophilia who have had large amounts of unsterilised blood products in the past have had hepatitis B and therefore have immunity. There is a vaccine against hepatits B which has been available since 1983. This should be administered subcutaneously to all patients with an inherited blood disorder including babies with haemophilia as soon as they are born.

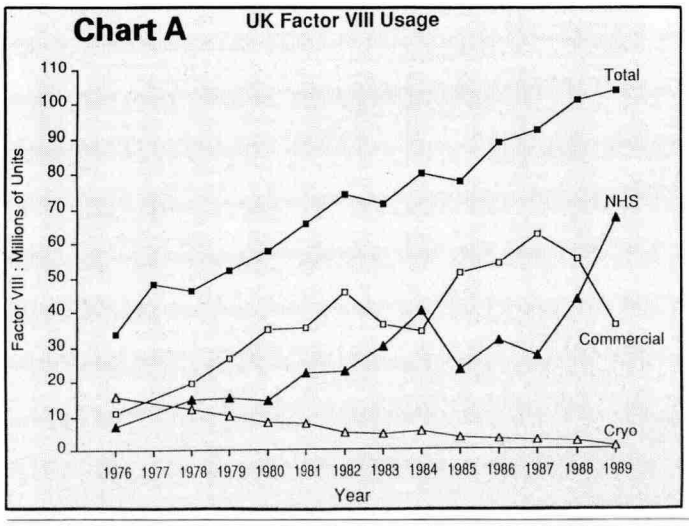


Christine Lee

Hepatitis D or the 'delta agent' only rarely caused hepatitis in people with haemophilia because it requires the presence of circulating hepatitis B virus to survive. It does, however, cause severe liver disease and should therefore be treated with interferon.

Hepatitis C is responsible for most of the hepatitis which occurs in haemophilia and it was formerly known as non-A non-B hepatitis. Since this virus was isolated just over two years ago large

Chart A UK Factor VIII Usage



Haemophilia and Hepatitis Past and Present

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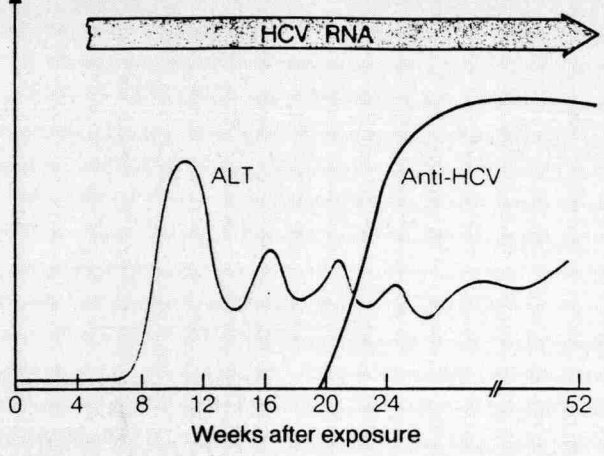
Chart B

Serological course in acute hepatitis C progressing to chronic hepatitis C



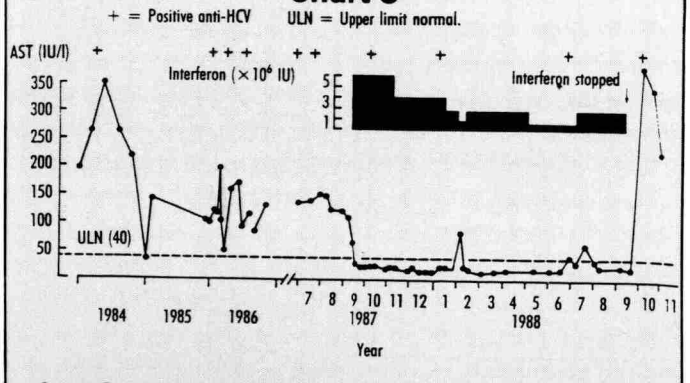
numbers of people with haemophilia have been shown to have antibody to part of the virus known as C-100. The rate of antibody in different populations throughout the world is 59%-85%.

BLOOD STUDIES

Studies on stored blood specimens from patients with haemophilia have shown that following a first infusion of factor VIII or IX the liver function tests become abnormal. There are enzymes released into the blood stream when liver cells, which harbour the virus die. This causes the "hepatitis" which is only rarely symptomatic. There then follows a period of chronic abnormality of the liver enzymes (transaminases) over a period of years. These commonly go up and down — the so-called "yo-yo" effect. The antibody to hepatitis C becomes positive. There is now more than one antibody which can be measured to C-100, C-22 and C-23. It is thought that the presence of antibody denotes presence of virus.

Hepatitis C seems to be a very slow progressive disease. In individuals who do go on to develop liver damage this is a very rare event and may take decades. In order to prevent such progression, treatment with the drug interferon is being investigated.

Chart C



Interferon treatment for chronic post-transfusion non-A, non-B hepatitis. Reproduced with kind permission of the British Journal of Haematology from Lee C A, Kernoff P B A et al, Br J Haematol, 1989, 72, 235.

Interferon is a natural product of "cytokine" which is released when viruses enter a cell. It can be produced on a commercial scale by infecting human cells in a culture with a virus and collecting the virus. It was first used to treat hepatitis C in 1986 when it was found that subcutaneous injections resulted in the liver enzymes becoming normal. Investigators felt that this represented the virus being prevented from multiplying. A number of studies are now being conducted, some in people with haemophilia, to work out questions of when to introduce the treatment, what dose to give, how often and how long. As yet, it is not a routine treatment, but within the next few years this is likely.

SIDE EFFECTS

Interferon, in common with most treatments, has side effects: these are "flu-like" symptoms and sometimes a lowering of the white cell count.

Liver transplants have now been performed in about ten people with haemophilia world wide. This may be necessary in the rare event of severe liver damage. Liver transplant "cures" haemophilia because factor VIII is synthesised in the liver. Thus once the operation has been performed and the liver is working, no further factor VIII treatment is needed.

THE FUTURE

There is no future for haemophilia and hepatitis. In our own unit we have 20 children under the age of 10 who have only received sterilised factor VIII or IX — mostly NHS 8Y or 9A. All are anti-HCV negative with normal liver enzymes and they are free from joint deformity. All are anti-HIV negative. We can be confident that children with haemophilia, who are treated with sterilised blood products and immunised against hepatitis B, will not develop hepatitis as a result of blood product therapy.

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TRAVELLING ABROAD?

We are now advised by the US Embassy that waivers will be issued to those who are HIV positive and travelling to the USA on holiday. The situation may improve further — always provided of course that enough people wrote as outlined in UPDATE!!

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Mum's Page

GRO-D

Injections are a 'fact of life' for people with haemophilia and their families but how have you coped and what are your experiences?

Here are some of our experiences and feelings.

● *'My name is GRO-A I am 26 and have two children, GRO-A 3 and GRO-A 9 months. GRO-A has haemophilia. At first I wondered how I would cope but by sharing experiences and learning I am coming to terms with it. He has his share of problems but copes wonderfully. I have been treating GRO-A at home for 6 months — when his veins allow it! However, even when I take him off prophylaxis to rest his veins I know if he bleeds I can treat him quickly and avoid any disruption to our lives.'*

● **'My son has been on home treatment for just over a year. He doesn't have very many bleeds and so I do not get regular practice! This week I had to do two injections on consecutive days and they could not have been more different. After a few tears he agreed to be co-operative on day one and I got to his good vein first time. HOORAY. Day 2 saw a very good boy doing his own local anaesthetic and all ready, and I missed and missed and it got more and more difficult as tension and emotions ran high with both of us crying. He bounced up ready to play, I was shattered! It isn't always easy but it's so good when it all comes together and so much better than a long trip to the hospital —we'll keep persevering!'**

● *'Following the shock of knowing our 10 month old son had severe haemophilia, life was extremely tense, wondering if each fall would lead to a visit to the hospital. At three and a half years old GRO-A was put on a prophylactic regime, which meant he had an injection of factor VIII three times a week to prevent the problems with bleeding episodes, and we soon learnt to inject at home. The last five years on prophylaxis have been wonderful. GRO-A has few problems, is able to enjoy school with no disruption as a normal eight year old and life is without tension.'*

'Can you imagine in your wildest dreams that your son will say 'Please mum, can I do my own needle today?' Well, that's what happened to me on Saturday 25.6.91. I couldn't believe my ears. My son, who is now eight, has been on prophylaxis since he was very small and although he now tolerates his treatment — only in his favourite vein! — there were times when he used to go crazy.

He very carefully inserted the needle and after a couple of tries passed it back to me. The next time he was due he said 'I know exactly what I did wrong the other day, now I'll get it right.' Sure enough he did and has done ever since. It's such a leap forward, a sign of maturity and a blessing to us. Now he can look forward to a weekend away with his grandparents and friends knowing he's totally in control of his own wellbeing.'

● **'When my eldest son GRO-A was two months old he was admitted to hospital with a swollen knee. After tests I was told that he has haemophilia. If I'm honest I must say that I found it hard to come to terms with. Three years later my son GRO-A was born. Tests showed that he too has haemophilia. I began to feel the need to talk and so when I was invited to meet a group of the other mums of children with haemophilia I jumped at the chance. It helped me tremendously. I no longer feel alone and more importantly I have come to terms with my sons' haemophilia.'**

We have all been helped in East Kent by our friendship. We hope Mum's Page will help you. Please write to us with your news and views so we can extend our page throughout the membership.

GRO-A

GRO-A — ***totally in control of his own wellbeing.***

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EMINENT GATHERING

Eminent speakers from around the world met at the Royal Society of Medicine in London recently to discuss the future of haemophilia treatment.

Significant advances in the purity and potency of blood products since the early 1980s formed the basis of discussion for the future of prophylactic treatment for people with haemophilia.

The session thoroughly endorsed the efficacy of high purity products and lent its weight to the need to encourage prophylaxis.

Commenting on the event, David Watters, General Secretary of the Society, was delighted with the views and opinions put forward.

"The Society has endorsed prophylaxis for some time and it is heartening to see such an eminent gathering adding its weight to the issue."

The session proceedings, entitled 'Factor VIII: Purity and Prophylaxis' has now been published and is available from The Haemophilia Society.

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