Witness Name: Katherine Victoria Burt

Statement No: WITN6392001

Exhibits: WITN6392002 - WITN6392267

INFECTED BLOOD INQUIRY

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The Bulletin

HEPATITIS C CAMPAIGN NEWS UPDATE

The hepatitis C campaign has been successful in continuing its momentum during the summer.

There has been a constant flow of items in the press and other media, where the aims of the campaign have been mentioned and the lack of action by the government highlighted.

This is the result of a number of factors, not least of which have been the actions of members approaching their own local media to speak about hepatitis C and how it affects them.

The recent product recalls by Alpha and BPL have also brought the issue of product safety into the public eye, as has the announcement that the NBA has started screening for Creutzfeld Jacob Disease among blood donors. These events have enabled the Society to highlight the need for the safest possible clotting factors to be used and to urge the introduction of recombinant clotting factors that are produced without using human plasma.

The issue of recombinant products and VAT imposed on them has attracted considerable media attention, and the pressure is mounting for their widespread use, which is one of the aims of the hepatitis C campaign.

Parliament is currently in recess, and there has been no response to the proposal put by the Society and Manor House Group to the Government for a £20 million hardship fund and £10,000 across the board payment for people with haemophilia infected with HCV. However, the constant mention of the campaign in the regional and national media will have helped to make the Government aware that the Haemophilia Society and Manor House Group are not letting the matter rest and will continue to press for Government action.

The Society will be pressing the Health Minister for action when the summer recess is over. In the meantime, it is important that people who have established contacts with their local media continue to use them to keep the issue in the public eye.

It is also important that members keep up the pressure on their MPs and the Government. Write to your MP asking why the Government hasn't responded to the Society's proposal for financial help. Ask your MP to write to John Horam in the Department of Health or the Prime Minister to ask why there has been no action.

If we can maintain pressure on the Government through MPs and the media we can keep the issue alive and encourage a speedy response.

Keep up the good work!

PHILIP MORRIS AWARD

GRO-A

Pictured is GRO-A receiving the Philip Morris Award cheque for £400, from Kevin Morris. GRO-A is studying timpani and percussion at the Guildhall School of Music. He has played with the All Souls Orchestra in London and at the Royal Albert Hall and has travelled abroad many times with the Oxfordshire County Youth Orchestra.

He is registered at the Oxford Haemophilia Centre and his application for the award was supported by Dr Paul Giangrande. GRO-A said he was "careful" and "aware" of the impact upon joints whilst playing percussion instruments but added that it did not dampen his enthusiasm.

Kevin Morris was impressed by GRO-A's commitment and obvious talent, he commented that the award reflected his fathers love of music and wished GRO-A the best for his chosen career. The award took place with Tony Wilson at the Society's national office. GRO-A is using the money to buy his own percussion instruments.

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EDITORIAL by Bulletin Editor Andy Cowe

The issue of product safety and the provision of the best treatment available is one that is very close to the hearts of everyone with haemophilia.

In Manchester, parents have been campaigning hard to obtain treatment for their children with recombinant factor VIII because they believe it is the safest product available.

So far they have had some notable successes. They have obtained national media coverage for their cause on numerous occasions and they have met with the executives of their hospital and centre director to demand an explanation for the lack of use of recombinant. Seven children at Manchester are now being given recombinant.

The hospital puts the blame on the health authorities responsible for purchasing treatment. The health authorities say that they are awaiting the Haemophilia Centre Directors' Guidelines on the use of recombinant. The delay, for whatever cause, in introducing recombinant is causing anguish among parents and children alike.

Surely, now would be an opportune time for the Government to centrally fund the use of recombinant so that its cost would not be an issue for the purchasing authorities and it would then be automatically available.

Meanwhile, the parents of Manchester fight on. Their determination is an example to all of us at the Society, and they deserve our support. If you want your child to receive recombinant factor VIII contact your centre to ask if it is available, and if not why not. The greater the pressure for the use of recombinant nationally. the more likely we are to obtain central funding from the government for this safer product.

Publications and Services available from the Haemophilia Society

Publications

The Society produces the range of books, booklets and leaflets listed below to help people with haemophilia.

- ★ Introduction to Haemophilia
- ★ Joint Care and Exercises
- ★ The Essentials of Haemophilia Care
- ★ Teaching Children with Bleeding Disorders
- ★ Haemophilia and Hepatitis C
- ★ Children's Haemophilia Book
- ★ Will making guide
- ★ NHSME Patient Perspective Booklet
- ★ Past copies of the Bulletin

Services

The Society works to help people with haemophilia from its national office and also via the local Groups. The services currently available from the national office are:

- ★ Information and help with benefits, in particular Disability Living Allowance
- ★ Hardship grants
- ★ Centeon Call service
- ★ Caravan Holidays
- ★ Adventure Holidays for children
- ★ Fund-raising support
- ★ Assistance with media enquiries
- ★ Information on treatments and blood products
- ★ Travel insurance advice
- ★ Information on travel regulations/ restrictions
- ★ Haemophilia Days and Family Days
- ★ One-off meetings on specific issues, such as hepatitis.
- ★ Parent Suport Network

For further information about the above services, or to check on the availability of Society publications, please contact the national office.

In this issue

RECOMBINANT FACTOR VIII AND PRODUCT SAFETY

Over recent weeks there have been a number of media items about the safety of plasma based clotting factor concentrates prompted by the recall of two products, one from Alpha and one from BPL.

The Alpha product was recalled from Manchester Children's Hospital following three cases of hepatitis A at the hospital, while the BPL product was recalled when it was found that the pooled plasma from which it was produced contained the hepatitis C virus.

Both recalls caused concern among the haemophilia community and prompted calls for the widespread introduction of synthetic (recombinant) clotting factor rather than plasma derived products. This was particularly marked in the Manchester region, where parents have been campaigning hard among their health authorities for its introduction.

The Haemophilia Society is strongly in favour of the use of recombinant products, because they should carry less risk than products that are derived from human blood.

Graham Barker, the Society's Director of Services commented that using recombinant minimises the risk of as yet unknown blood borne viruses infecting people with haemophilia.

"While clotting factor concentrates are produced using human plasma there will always be some risk, however slight, of blood borne viruses getting through," said Mr Barker. "This is particularly true for as yet unknown viruses. If an unknown virus is currently present, there is no way of testing whether the viral inactivation techniques used on plasma products are effective and so there is always the risk of transmission. We need to get human plasma out of the chain, so that people with haemophilia can receive their treatment confident that the product they are using is as safe as possible."

The Society would like to hear from people who are trying to obtain treatment with recombinant products. Please contact Graham Barker at the national office and let us know if you have had any success, or whether it has been refused and if so on what grounds.

HEPATITIS WORKER DEBORAH GILLESPIE

Deborah Gillespie started work with the Society as hepatitis worker on 29 July this year.

She joined the society from the British Diabetic Association where she worked for the last four and a half years in the Services Division.

Her previous experience includes several positions providing support for people with diabetes, including addressing the needs of adults, children and young people, parents, carers, partners and health care professionals; operating a telephone helpline; provision of benefit information; insurance information and general support on all aspects of living with diabetes.

"I'm looking forward to learning about and addressing the needs of people with Haemophilia and Hepatitis," she said. "The Haemophilia and Hepatitis C Research Report has provided a very interesting insight into the needs of those affected by HCV. I intend to act on the recommendations of the report and I would very much welcome calls from people affected by the virus if they would like to share their experiences."

To increase the opportunity for individuals to telephone the Society, Deborah will be operating an "out of hours" service from 30 September on a Monday evening from 5.00 pm to 7.30 pm. This will hopefully provide a more convenient time for some callers to reach the Society.

She is looking forward to receiving calls and listening to ideas on how the Society can best support people with Hepatitis and others affected by the virus.

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THE RED RIBBON PAGE

HIV PROTEASE INHIBITORS

by GRO-A

In America, the government body dealing with drugs (FDA) has learned of approximately 15 case reports of spontaneous bleeding episodes in HIV positive patients with haemophilia who were being treated with HIV protease inhibitors at the time of the event.

Of the cases of spontaneous bleeding preliminarily associated with these drugs , 11 have involved haematomas and 5-6 haemarthroses (one patient reporting both).

None involved serious injury or death. The majority of patients have been able to continue taking their protease inhibitor therapy in spite of the bleeding event.

Up to now, there is no conclusive evidence to establish that the protease inhibitors are the cause of these spontaneous bleeding episodes.

However, as these products were all approved in the States under the government's accelerated approval mechanisms for treatments for life-threatening illness, the US government believes it is important to investigate and question about safety that arises early in the marketing experience.

There are three HIV protease inhibitors currently approved for marketing in the United States for the treatment of HIV infection: saquinavir (Invirase) from Hoffman-LaRoche, ritonavir (Norvir) from Abbott Laboratories, and indinavir (Crixivan) from Merck Research Laboratories.

To date all cases reported have involved European patients. At this time it is unknown whether there is a causal relationship between the use of protease inhibitors and bleeding episodes in patients with haemophilia.

However, in light of these reports, the drug manufacturers recommend that centres need to monitor patients with haemophilia for spontaneous bleeding episodes whenever any of the protease inhibitors are used as part of treatment for HIV.

Patients with haemophilia and HIV infection who are currently using protease inhibitors should not discontinue using their treatment but should instead consult with their treatment centres if they have any concerns.

PROGRESS FOR BIRCHGROVE WOODLAND PROJECT

The Birchgrove Woodland Project is currently progressing well. Through the generosity of the members of the Haemophilia Society we have received many tree planting sponsorships.

Recently we have not been able to issue certificates and acknowledgements as some people have omitted to provide us with their return address. Anyone who has not yet received their certificate of sponsorship should contact us as soon as possible with the address where they require it be sent.

NEW ADDRESS FOR BIRCHGROVE GROUP

The Birchgrove Group has a new address. It is:

The Birchgrove Group
PO Box 9, Abertillery, Gwent NP3 1YD
Tel: 01223 373560 (Admin)
GRO-C

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REPORT ON THE WORLD FEDERATION OF HEMOPHILIA CONGRESS - JUNE 1996

The World Federation of Hemophilia Congress took place in Dublin earlier in the summer. Here we print the impressions of three clinicians who attended.

Dr Elizabeth Mayne
Belfast Haemophilia Centre

The sprawling landscaped Belfield site of Dublin's University College sprang into life on Sunday, 23rd June 1996. The multiple grey buildings and the new O'Reilly Hall were transformed into a hiving, colourful complex, thronging with delegates from 75 countries who, during the ensuing days, were to bask by the lake in the warm summer sunshine in between the scientific sessions, the posters and the endless socialising. The 22nd Congress of the World Federation of Hemophilia had begun.

The statue of Molly Malone, a well known Dublin landmark

The opening ceremony was memorable. The speeches were refreshingly succinct and to the point. The following musical entertainment was heart-stirring, both feet and hands tapped and clapped appropriately. The promising start was confirmed by the next four days of hectic activity. During the first Plenary session, haemophilia care in India was highlighted, or perhaps more poignantly, its lack was emphasised. Dr Chandry described the situation with great clarity. He indicated the irrelevance of the conference motto "Care to Cure" for that continent and probably to much of the developing world. It is clear that so much has been achieved but on a world wide basis there remains so much still to be done. His lecture provided real food for thought and can be considered a true highlight of the entire meeting.

In stark contrast, a further highlight was indeed the gene therapy session. It was attended to overflowing. All aspects of the subject were presented with lively discussion and questions and answers. It is clear that all answers are not yet available and we look to the future for more progress in this field.

There was a plethora of lectures, presentations and posters. Perhaps the latter were not all displayed to full advantage as in one poster area the light was of a dim, religious twilight type. However, such a criticism is minor when one considers the opportunities for discussion, the exchange of problems, comparisons of management with patients, colleagues and carers of all disciplines associated with haemophilia management. Equally well one must highlight the pharmaceutical companies who afforded all delegates the opportunities for questions regarding the various blood products.

GRO-D

A lighthearted moment for (standing l-r) Terkel Andersen, Jenny Ross, Peter Levine (Chairman) Alexandros Vargas and Ashok Verma

One must mention with acclaim one company's puppet show, another's T-shirts and overall the warm hospitality of all concerned.

The Thursday morning was special as the conference was honoured by a memorable visit by President Mary Robinson. She spoke with warmth, humour and much insight into the nature of and the background to the conference and the problems which it was addressing. Afterwards she mingled with delegates during the coffee interval. Many were afforded the opportunity of having a few words with her. In the past, it has been custom for all the prominent or famous individuals of the host country to make a brief appearance at the opening ceremony. However, in Dublin the reverse was practised and it was indeed a very successful and memorable occasion. No comments on the conference would be complete without a mention of the warmth and hospitality of the hosts and their unforgettable kindness to all. One mentions in particular the official reception in the splendour of Dublin Castle, the conference dinner and then the many other events, whether inside or outside Dublin City. Warmest congratulations to Dublin on the success of a truly great conference.

Dr Mark Winter
Canterbury Haemophilia Society

The prospect of gene therapy is a tantalising one for patients with haemophilia, and all who look after them. In many ways, haemophilia is an ideal disorder to be treated by gene therapy because many of the problems that would have to be overcome for gene therapy to be applied to other illnesses are not present. For instance, gene therapy for patients with diabetes would require a precise and exact amount of insulin to be delivered at a constant rate so that the blood sugar became within the normal range and was neither too low nor too high. In haemophilia, no such controls are really required because there are no dangers associated with high levels of factor VIII. Furthermore, experience of treating patients with prophylaxis tells us that it is not necessary to make their levels of factor VIII (or factor IX) normal for them to be free of spontaneous episodes of bleeding. Indeed, people who are born with factor VIII levels above 5% tend to bleed spontaneously extremely rarely.

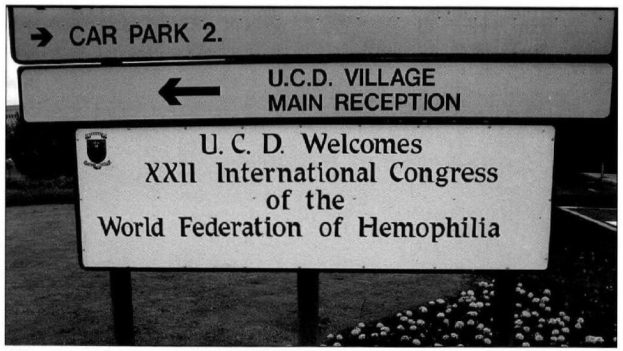
In other words, if gene therapy could be given to patients with haemophilia, even the modest increase in the basic level of factor VIII or IX to say 5-6% might prevent nearly all episodes of spontaneous bleeding.

The principle of gene therapy involves the insertion of the factor VIII or factor IX gene into a suitable carrier or vector. Ideally the factor VIII gene becomes inserted into the DNA of the vector. The vector is then inserted into an appropriate cell line which might consist of cells taken from the bone marrow, or the lining of the blood vessel or the liver. Once the package has been prepared it can be implanted into the patient (the exact means how this might happen has not been identified) and would release factor VIII or IX directly into the circulation of the blood. It looks as if the packages of gene therapy would not last forever but would need replacing every few months or so. One of the major roles of the Haemophilia Centre would be to monitor the levels of factor VIII and factor IX to assess whether a replacement package of gene therapy would be required.

The session on gene therapy in Dublin was particularly interesting because there are clearly a number of developments that are taking place that are of considerable interest. At the same time, it is apparent that a number of formidable obstacles remain before we can expect to see the introduction of gene therapy. Nevertheless, the situation is rather similar to that which existed with the introduction of recombinant factor VIII concentrates in that it looks increasingly possible that gene therapy can be shown to work in animals, and it is therefore a question of fine tuning experiments that have been shown to be effective.

It is hard to say at this time when we can expect to see gene therapy introduced. It looks likely that gene therapy will be available for patients with factor IX deficiency first of all, because factor IX is relatively smaller than factor VIII. Quite recently, two boys with factor IX deficiency have been treated with gene therapy in China although the treatment did not prove to be effective.

When it does arrive, gene therapy is going to cause a dramatic change in the lives of patients with haemophilia. In the words of one of the speakers in Dublin it should prove to be "efficient, safe and convenient".



Signposts point the way to the XXII Congress

Dr Angela Thomas
Consultant Paediatric Haemotologist, Edinburgh

Delegates to the International Congress of the World Federation of Hemophilia, an eclectic mix of patients, carers and scientists, gathered in Dublin for the 22nd meeting in June this year.

The programme was full and sessions often simultaneous but certain areas were of particular interest to me, one such being recombinant factor VIII therapy. There were several sessions devoted to this theme. Encouragingly, world-wide experience has shown that its efficacy and safety, particularly with respect to haemostatic properties and inhibitor development, are very good, but there are obvious problems about cost. Two of the products are now licensed, with a third under development.

There were sessions on prophylaxis and it was quite enlightening to hear that the Swedish who pioneered prophylaxis in children and teach mothers to give the prophylaxis from a very early age in fact have a learning programme which extends over a year. I had always assumed that proficiency was achieved in a much shorter time and feel heartened that we are not too far behind.

I also met a doctor, Tatiana Andreeva, from St Petersburg who has to cope with more than a thousand people with haemophilia and whose lab routinely runs between 8-10 diagnostic factor VIII levels a day. There is virtually no factor VIII concentrate, most of the patients being treated with cryoprecipitate. A very large geographical area is covered and most patients are educated about how to avoid bleeds or how to cope with ones that they have but there is no possibility of arranging treatment for most of these people. The factor VIII concentrate that is available is mainly used to cover orthopaedic operations for those with the most severe joint damage. One of the first aims is to try to virally inactivate cryoprecipitate rather than to generate factor VIII concentrate.

Near the end of the conference, Mary Robinson, President of the Irish Republic, gave us all a lovely welcome - very Irish - and spent time talking most particularly to those people with haemophilia and their carers who had travelled thousands of miles to attend. She spoke warmly of progress made in haemophilia care and stressed the importance of continuing this work. When I left Dublin, I felt encouraged that the progress made will continue through the efforts and co-operation of all those involved with haemophilia.

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A LIVER TRANSPLANT THAT CHANGED A LIFE

This article, which was sent to the Bulletin by the wife of one of our readers graphically demonstrates the dramatic improvement in the quality of life that can be achieved by a liver transplant.

The writer is happy to talk to anyone who would like to know more about the practicalities of coping with a relative having a transplant at the Liver Unit of the Queen Elizabeth Hospital in Birmingham. The name and phone number are available through the Haemophilia Society national office.

"If you're not ill enough to see the GP then do the washing up," I growled to my wheezing husband. So it was at the end of November GRO-A who had severe haemophilia B and hepatitis C, finished work, went to the supermarket and then to make an appointment. At the surgery the receptionist listened to his wheezing. "The doctor will see you now," she insisted. Within 2 hours GRO-A had been admitted as an emergency to our local hospital. Fluid collected in his abdomen and in his lungs was stopping his heart and lungs from working.

At first we did not take what was happening at all seriously. As far as we knew GRO-A's liver damage was only mild - liver function tests two months previously had been stable and not at all alarming. He had suddenly put on weight and girth and become breathless but nothing else. Within three days, however, we learned that his liver had started to give up the ghost. The consultant was quite clear; GRO-A life expectancy was very limited and the only thing that could possibly save him was a liver transplant and he thought he needed one fairly urgently. He referred us to the Liver Unit at Queen Elizabeth Hospital Birmingham - one of the biggest liver transplant centres in the world, with a wonderful Haemophilia centre on tap.

After a preliminary interview at Birmingham we had to wait 6 weeks for admittance and in that time GRO-A became less and less mentally acute, very weak and aged twenty years. By the time he was admitted for assessment - to see whether transplant was possible - he was so weak that every test seemed to send him into crisis. The assessment which should last 10 days lasted three weeks. At the end he was thought to be too ill to return home before the transplant which, despite everything, they felt they could do. Having seen him near death after a small liver biopsy I asked how he could possibly survive transplant. "Our job is to cope with all the problems liver failure throws at us, not to be overcome by them," I was assured. I learned the first Liver Unit lesson; people who are very, very ill can be brought back not just to life but to near normality. Time and again I saw other patients in extremis one day and sitting up the next chatting to their neighbours. This coupled with the rule of life of people with haemophilia - not to worry about the "what ifs" but only to worry when you really have to - helped us to manage the stress.

The other thing that helped us through was the loving care and attention of all the staff and in particular the haemophilia nurses and doctors. Their constant presence and the complete understanding of the Liver Unit that the haemophilia experts were to be consulted at every stage were immensely reassuring.

> "Our job is to cope with all the problems liver failure throws at us, not to be overcome by them"

GRO-A was put near the top of the urgent list in the middle of February this year. The terrible shortage of organs however meant that he did not receive a transplant until early June. In the intervening time he recovered sufficiently to come home and we had three months of short periods of calm at home followed by mad and very fast ambulance dashes the 125 miles back to Birmingham as crisis followed crisis. It was a difficult time and more and more we were aware that not everyone survives the wait for the right organ. It was also very hard financially; obviously he could not work and being self employed had only limited benefits. I was torn between needing to bread-win and needing to nurse him. His crises could come on in 5 minutes leaving him too weak even to phone me so every hour away from home was a nightmare but I could not afford to give up work altogether.

At length the phone call came - at 9.30pm. We shook for ten minutes and then left for the long drive up the M5. We arrived at the hospital at 2am; Ian's operation started at 2.30 that afternoon. Most of the intervening time was spent in careful blood clotting measurement and massive infusion of factor IX, platelets and fresh frozen plasma. By 10.15pm he was tucked up in Intensive Care to start recovery.

Recovery was inevitably uncomfortable for a couple of days but GRO-A was soon over the worst and then made rapid and astonishing recovery. One of the huge boosts to morale was the knowledge that he no longer had haemophilia and the delight over this seemed to get to everyone in the hospital. Bitten lips, which used to drip blood for weeks, stopped after 30 seconds - and this became a matter of rejoicing not just for us but for everyone, from consultants to porters. While the haemophilia family is a supportive and warm one to belong to the ex-haemophilia club is pretty wonderful!

GRO-A was home three weeks after the operation and he is doing very well indeed. It is only 6 weeks (at the time of writing) since the operation and things can go wrong but there appears to be no reason at present why they should. Our life is already better than it has been for a few years. Neither of us had realised how insidiously a badly functioning liver affects you and now he has a good one he has a sparkle back which he had completely lost. He still has hepatitis C of course but we are hoping that this time round it does not have such a devastating effect on him.

GRO-A was lucky; his consultant in our local hospital thought it was worth looking at transplant despite the complication of haemophilia (and as it happened haemophilia did not seem to cause huge problems at all during the actual transplant - and after it of course there was no haemophilia to worry about.) Organ shortage puts very real limits on the numbers who can be given transplants (so encourage your friends and family to carry donor cards) but there does not seem to be any reason why people with haemophilia should not be given an equal chance of life through transplant.

Editor's note for more information about liver transplants see the Dear Doctor section on Page 12 of this Bulletin.

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WHY IS HEPATITIS C ALL IN THE MIND?

This article was written by a member of the Manor House Group and any comments should be addressed to: Manor House Group, PO Box 128, Nantwich, CW5 8PQ. We would especially like to hear the views of medical professionals.

I would like to challenge the historical medical viewpoint of both HIV and HCV, and in the case of the co-infection the interactive relationship of the two viruses.

I have become convinced that had our friends with HIV not been co-infected many may still be with us. I became convinced when I read an article in the Birchgrove magazine where the author was basically listing the symptoms he was suffering as a result of his HIV infection. What surprised me was that they were, to a large extent, the same as my own and I am only hepatitis C positive.

Being HIV negative I would not have had the audacity to begin to think this way without the support of several friends, now sadly departed, who were co-infected but eventually succumbed to the effects of HCV. I have the knowledge that they all agreed to my thesis.

We know HIV needs a catalyst, another virus or additional action upon the immune system. In many cases outside of the haemophilia community I believe it is often drugs which activate the virus, but what about the haemophilia community who, by and large , do not use drugs; what activates the virus? I began by wondering if it could be the continued use of factor VIII (or IX) itself, but could it be the hepatitis C virus? Could we have lost many friends with liver problems whose death has been attributed to HIV infection when, had it not been for their co-infection with hepatitis C, they may still be with us? Have the medical profession, as I believe they have, failed to understand the full implications of the hepatitis C virus?

I appreciate the whole issue is far too complex to give a yes or no answer especially as all our bodies fight infections in different ways, however this in itself could be the reason my thesis works; not everyone with HIV has succumbed to the most commonly known HIV related illnesses, haemophiliacs have not suffered from the same illnesses as, say, the homosexual community, so why could hepatitis C not have played a part.

I would like to stress to all my friends with HIV that I do not in any way wish to understate the seriousness of HIV itself, I simply wish to question what effect HCV has had on our health, find out whether the effects of HCV exacerbate (as they must) the effect of HIV on the immune system, and why people who have HCV ONLY have similar symptoms and health problems as those who are co-infected. I would like to challenge the medical thinking from the people who have spent many years researching blood borne viruses to consider once again the effect the hepatitis C virus has on the WHOLE body because I cannot understand why HIV negative members of the Manor House Group suffer similar symptoms to those which are more generally attributed to HIV. The Group is aware of the fact that for over fifteen years HIV negative but HCV positive patients have consulted various doctors only to be told by many that their perceived symptoms are all in the mind and are probably caused by depression. We know they are not.

I would like to know how many people with haemophilia have - since 1985 - died from various causes but have during a post mortem been found to have a seriously damaged liver which, had it not been for those causes, would more than likely have died as a result of liver problems. Have the implications of HIV distorted the true mortality rate from hepatitis C infection?

DEAR DOCTOR...

I have hepatitis C and severe liver disease, what are the chances of my getting a liver transplant and what is the experience of liver transplant for people with haemophilia?

The first thing to clarify is what you mean by severe liver disease. Severity can not usually be assessed by the routine blood tests you have when you visit your centre. There are two ways of assessing severity:

a) Through a liver biopsy. Although the finding of cirrhosis implies severe liver disease, the progression from this to liver failure is not always rapid. Two of our haemophilia patients who were shown to have cirrhosis on liver biopsy, did not require a liver transplant until 12 years later. It is not always essential to perform a liver biopsy and some haemophilia centres do not do them.

b) Clinical examination and special investigations. Liver failure can be detected by clinical examination (eg swelling of the abdomen due to fluid), by the finding of swollen veins on the inside of the oesophagus (gullet) on endoscopy, or by tests that look at the liver's capacity to manufacture proteins.

Once a patient with haemophilia has been found to have advanced liver disease (liver failure) he should be referred to a liver transplant centre for assessment. There are at least 7 liver transplant centres in the UK and 2 of these have experience in transplanting people with haemophilia. Different centres have different criteria for performing a liver transplant but these are usually similar, centring around the presence of advanced liver failure and life expectancy of less than one year. People with haemophilia have the same access to transplantation as anybody else. It is our policy to refer all patients with liver failure for assessment by a liver transplant team. In general, only patients younger than 65 years will be considered, and in haemophilia the patients must not have an inhibitor. It is well known that alcohol accelerates the progression of hepatitis C and it is unlikely that persons with high alcohol intake and hepatitis C will be transplanted unless they show that they have stopped drinking.

As far as I am aware there have been 7 liver transplants of people with haemophilia in the UK and around 30 others world-wide. In patients who are HIV negative, the results are at least as good as those in non-haemophiliacs where survival 5 years after transplantation is around 80%, despite the fact that the new liver gets re-infected with hepatitis C in every case. The liver manufactures all the clotting factors including VIII and IX, so once the new liver is in, the haemophilia is effectively cured and patients no longer need treatment with clotting factor concentrates.

After a liver transplant patients have to take immunosuppressive drugs for life to prevent rejection of the new liver. This would be undesirable in HIV positive patients who are already immunosuppressed. Each case of liver failure in an HIV infected person with haemophilia has to be assessed on its own merits, but provided the CD4 count is stable and at a reasonable level (eg greater than 400), some but not all centres would consider liver transplantation.

Dr M Makris, Senior Lecturer,
Sheffield Haemophilia and Thrombosis Centre.

ABOUT VON WILLEBRAND'S DISEASE

by GRO-D

I keep hearing and reading about von Willebrand's disease being only a mild condition. I also hear and read that von Willebrand's disease affects 1% of the population, so it is a common condition.

However, it is a condition that affects both male and female and is classified into different types. Whereas the majority of sufferers are classified as "mild to moderate" or just plain "mild", a small number of those with the condition are severely affected. In its severe form, von Willebrand's disease can be nasty and unmanageable. Little has been written about the severe form where very prolonged bleeding occurs and joints and muscles can be affected. For severely affected females, problems with periods and internal bleeding may be huge, leading to anaemia and the need for transfusion.

Modern treatments, factor VIII replacing cryo-precipitate, tranexamic acid and DDVAP, all help towards controlling bleeding episodes although DDVAP is not used to treat severe Von Willebrand's disease. For those using factor VIII concentrates, the risk of contamination by viruses is as great as for sufferers from Haemophilia A and B.

Since I have lived for 22 years as the parent of a daughter with severe von Willebrand's disease, I do know something about the care and management of this condition from day to day. During these years, as I have become involved in conferences and meetings organised by the Haemophilia Society, I have met other sufferers, male and female, and their families. Tales of woe about von Willebrand's disease have differed but many have been traumatic.

It is good that we get together and share information. It is good that our specialist doctors and nurses and all those who care for our families share their knowledge and expertise with us. I am concerned, though, that many doctors, senior and junior, and general practitioners have little or no idea about von Willebrand's disease. They may be under the impression that "it's only a mild condition" and so do not need to concern themselves too much. I feel that the time has come to educate more people in how to deal with this alarming disorder. For, if approximately 1% of our population suffer from it in some form, surely it should be more widely known about and hence understood.

I have become a volunteer on the recently formed Parent Support Network. From my calls to families who have asked for help through the network, I find that families with children with von Willebrand's disease are feeling very isolated.

The Haemophilia Society will be suggesting a von Willebrand's disease "spot" at the forthcoming Chairman's Conference in November 23rd and 24th, where anyone can come and bring ideas as to how we might effectively begin this process. A booklet on von Willebrand's disease will be put together when the necessary funding can be found. Hopefully, some publicity about the problems that people have to face will make the medical profession at least more aware and in turn able to help those with this distressing condition, especially those with young children who are struggling to come to terms with it.

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HIV – Let's Talk

£8.00 per copy + £1 postage & packing.

It is difficult to talk about the pain of HIV infection, the illnesses, and the separations, and the bereavements. One of the most difficult tasks has to be talking about illness and death with children. We have to help those children manage their own reactions and manage our own.

We have used our personal experiences with families to illustrate ways to talk together and share concerns and thoughts. Everyday things like photographs, pens, crayons, paper, books, videos and even water have been used so that feelings can be shared whatever the age of the child.

The book has been written for both adults and children, and is designed to be used in several ways. Some families may feel able to work with the child themselves. Some may want the guidance of a professional, and others may wish the professional to undertake the work for them.

Enquiries for both booklets please to: Linda McBride, Regional Haemophilia centre, Royal Victoria Infirmary, Queen Victoria Road, Newcastle Upon Tyne, NE1 4LP

ONE WORLD - ONE HOPE

This is the message of this years World Aids day (WAD), but what is the point of this day to people not already living with HIV in their family? Why for example is this not on the red ribbon page that normally takes all the HIV articles?

One of the original ideas of WAD nearly 10 years ago was to try to bring HIV to the attention of people worldwide and to try to make people aware that even though it did not appear to be of any importance to them that they needed to know about HIV. This way it was hoped societies would show greater care and concern to those people who were ill.

The recent Hepatitis A cases in Manchester are a timely reminder to us all how important the safety of our treatment is, regardless of if we have HIV or not. The scandal involving infected blood products and HIV infection played out in almost every developed nation highlighted this.

However, it should not be seen to be all doom and gloom, the recent Vancouver conferences had much more hopeful views on treatments than in previous years.

World Aids Day is now a major international event designed to improve peoples understanding and strengthen the worldwide effort to prevent the spread of HIV.

It is co-ordinated internationally by the Joint United Nations Programme on HIV (UNAIDS) and adopts a theme each year. The theme this year is One World One Hope in an effort to highlight the similarities of everyone wherever they are in the world and to try and involve people and families that have not (yet) been touched by HIV. And that's why this is not on the Red Ribbon page.

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PATIENT SURVEY OF BRISTOL HAEMOPHILIA CENTRE

Last year the Bristol Haemophilia Centre commissioned the Patient Survey Unit of the United Bristol Healthcare (NHS) Trust to undertake a patient opinion survey to look at the service provided by the Centre.

A questionnaire was drawn up following discussions with the Centre staff and interviews were held with 12 randomly selected patients. The final questionnaire was sent to 104 patients and 74 replied - a response rate of 71%.

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The survey's primary focus was not on clinical matters but instead the practical issues affecting the patients, such as car parking facilities, the waiting area, what happened out of normal hours if you went to the Accident & Emergency Department, the provision of information and access to particular services such as physiotherapy, HIV and HCV.

A report was produced, and although the full report was not circulated to all patients, the major findings were presented at an evening meeting at the Centre to which all patients were invited. The author of the report explained how the survey was conducted and highlighted some of its main findings.

There was also a presentation by a representative of the Centre staff, Sister Mary Edgar. She highlighted the good and bad points coming out of the survey but also said how the Centre was going to respond to the views of the patients.

She spoke about what had already been changed (for example introducing a pre-clinic questionnaire and updating the haemophilia treatment protocols for Accident & Emergency); things that they were working on (such as the introduction of a dedicated computer system to "keep tabs" on patients and improve communication); and things they would like to do.

This last list included the provision of a new haematology centre, a full-time specialist nurse, the provision of parking for disabled people and the provision of social work support.

The meeting also included a presentation from a liver specialist on hepatitis C - an issue that the survey showed many patients felt they had received insufficient information about.

The patient audit and the presentation of its results at an open meeting for all patients has had a number of benefits. Clearly the Centre staff and hospital authorities are now much more aware of the patients' views on the services provided. Some changes have already been implemented and others will hopefully occur in the near future.

Holding the survey has produced a clear, common agenda for change that staff and patients can work towards. The Centre Director has proposed setting up a small group of patients and staff to see how things can be taken forward.

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NEW HAEMOPHILIA CENTRE DIRECTORY

The 1991 Haemophilia Directory has now been updated and published by Bio Products Laboratory, in collaboration with the Haemophilia Society.

The new directory gives a description of each UK haemophilia centre including its status (Comprehensive Care Centre or a Haemophilia Centre), contact numbers, names and opening times.

The hospitals are listed by NHS Region at the front and alphabetical order at the back, so you can easily locate any centre in the directory.

You will be able to collect your new copy of the Haemophilia Directory from the centres across the UK or alternatively from the Haemophilia Society.

RECOMBINANT FACTOR FOR SCOTLAND

We are delighted to report that the Secretary of State for Scotland has released £1.1 million to buy recombinant Factor VIII for use in Scotland. The money is being used by the Scottish National Blood Transfusion Service to buy recombinant products from two commercial producers. It is understood that the use of recombinant factor will be phased in gradually. This development is something the Society has been pressing for some years now.

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