Witness Name: Katherine Victoria Burt

Statement No: WITN6392001

Exhibits: WITN6392002 - WITN6392267

INFECTED BLOOD INQUIRY

WITN6392150

The Bulletin

1987 – No. 2

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

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

The Haemophilia Society
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NEW £55 MILLION BLOOD PRODUCTS MANUFACTURING UNIT OPENED

HRH The Duchess of Gloucester opened a new production unit at the Blood Products Laboratory at Elstree in Hertfordshire at the end of April, signalling excellent news for people with haemophilia.

The new unit, representing a commitment of £55 million by the Government, will use some of the most advanced techniques and equipment of its kind to process human plasma into a number of vital blood products – particularly Factor VIII. It has three times the processing capacity of the old plant on the site and will enable the whole of England and Wales to be self-sufficient in such products.

The Blood Products Laboratory (BPL) is managed by the Central Blood Laboratories Authority, which is a special health authority set up within the NHS back in 1982.

It is estimated that once the new laboratory is fully operational it will be able to make blood products worth up to £60 million a year for the National Health Service.

NEEDS

Limited production will start later this year and should provide England and Wales with 75 per cent of their needs as the new plant begins to become fully on stream. Self-sufficiency is now projected by 1989.

The BPL is provided with human plasma by the National Blood Transfusion Service and the new unit will process 450,000 litres of this annually.

Plasma is separated into a variety of components by BPL by

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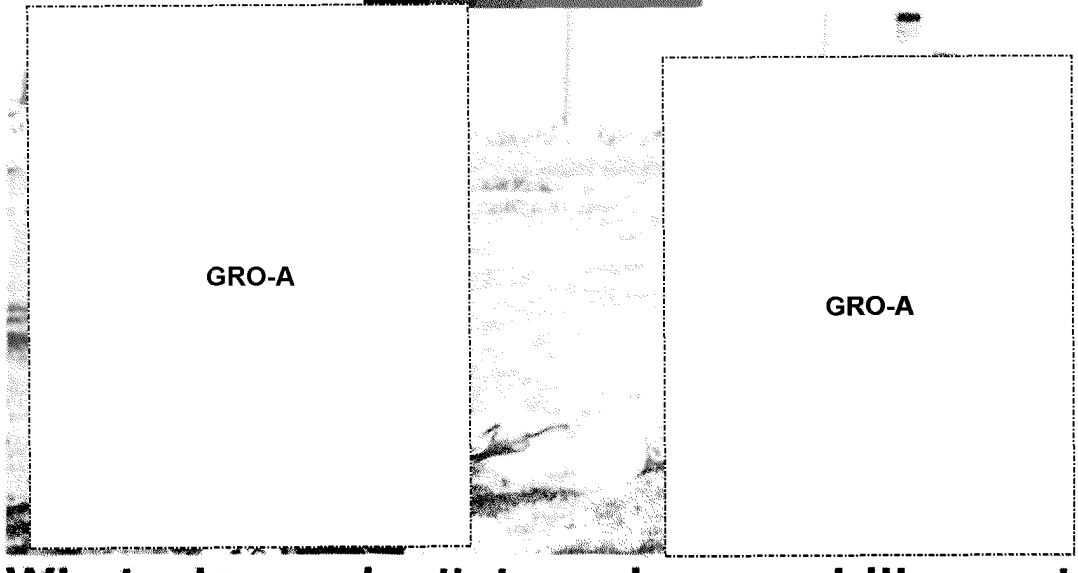
THE LONDON MARATHON

DR MARK WINTER, Director of Margate Haemophilia Centre, completed the 26.2 mile course in the amazing time of three hours twelve minutes.

We offer Mark our warmest congratulations on this achievement. Only a few weeks earlier he was to be seen hard at work, at our Annual Seminar in Newcastle, where he performed miracles with a baby's safety pin!!

Please collect your sponsorship monies as soon as possible and send them direct to: *Mr C Scott,* GRO-C GRO-C

A very warm *'thank you'*.



What does skydiving, haemophilia and these young people have in common?

Find out on page 13.

TRIBUTES

The haemophilia fraternity loses two of its finest men . . .

PROFESSOR R.G. MACFARLANE CBE, MA, MD, FRCP, FRS

Devoted his life to the care of haemophiliacs

PROFESSOR GWYN MACFARLANE, the President of the Haemophilia Society, died suddenly at his home in Scotland on 26th March, aged 79. We are very much aware that we have lost a good friend and supporter who had devoted his life to the care of people with haemophilia.

Professor Macfarlane's interest in haemophilia began to develop while he was an undergraduate, when he wrote his first paper on the subject, before going on to win the London University Gold Medal for his MD thesis on the haemostatic mechanism.

After that, he made significant contributions in many aspects of haematology but his international reputation was established through his work on blood coagulation. Some members of the Society still remember being treated by Professor Macfarlane when he was their doctor at St Bartholomew's Hospital, London, in the 1930s and introduced them to Russell's Viper Venom as a clotting agent.

★ *The late President of the Society, Prof. Gwyn Macfarlane.*

It was at Oxford, in partnership with Dr Rosemary Biggs, that he undertook his original work in determining the mechanism of blood coagulation. In the 1950s, the Churchill Hospital in Oxford became known throughout the world for its excellence as a centre for both scientific research and patient care, and their joint work *Human Blood Coagulation and its Disorders* is still the standard text-book on the subject.

Professor Macfarlane's academic achievements were recognised by his being elected a Fellow of the Royal Society in 1956. In 1959, the Medical Research Council established a blood coagulation research unit under his direction. He was elected to a Fellowship of All Souls in 1963 and appointed Professor of Clinical Pathology at Oxford in 1965.

He retired from active involvement in scientific research at the age of 60 and applied himself with characteristic enthusiasm to his new interest as a writer. He wrote two outstanding biographies, on Sir Howard Florey and Sir Alexander Fleming, giving a full account of the discovery of penicillin.

As far as the Society is concerned, Professor Macfarlane was involved in our activities from the very beginning, for it seems that it was a group of his own patients who first thought of meeting together to support each other and discuss their common concerns. From those early days, Professor Macfarlane continued to be actively concerned with the development of our work. In 1976 he agreed to the use of his name for **The R G Macfarlane Award,** the highest honour the Society is able to bestow on those who make outstanding contributions in the field of haemophilia. In 1983, Professor Macfarlane agreed to accept the appointment as President of the Society.

During the last few years Professor Macfarlane was greatly exercised by the advent of 'AIDS' and encouraged us to pursue ways in which those who were disadvantaged by HIV infection might receive special financial recompense from the Government. Shortly before his death, he was still considering plans for helping people with haemophilia to present their case for such special consideration.

A Memorial Service was held in the Priory Church of St Bartholomew the Great, Smithfield, London, on 9th June, when members of the Executive Committee were present, remembering his work as a medical scientist and the way in which he cared for people with haemophilia as the President of our Society.

FRANK SCHNABEL President of the World Federation of Hemophilia

'A Man of Vision and Dedication'

Members of the Society who have attended Congresses of the World Federation of Hemophilia will know how much we owe to FRANK SCHNABEL for his pioneer work in bringing together the National Organisations for such international meetings. His death from heart failure on Friday, 1st May, marked the end of that long period during which he led the World Federation with such vision and dedication.

At the first meeting in 1963, six member organisations accepted Frank Schnabel's invitation to join him in Copenhagen. There are now 64 countries represented in the Federation and that development is itself a tribute to the leadership he exercised for almost 25 years.

The story of Frank Schnabel's life is one of outstanding achievement and of triumph over adversity. When he was a boy, there was little to be done for people with haemophilia. He was so affected himself that his legs were supported by calipers. However, he was so determined to overcome his handicap that he worked his way through school, then university, and qualified to work as a financial analyst. Eventually, he set aside his calipers and began to walk unaided and to travel throughout the world in the cause of haemophilia.

At the World Congresses, he was tireless in encouraging all who came to him for advice and to discuss plans for the development of their own national societies at home. He was particularly attentive to those who came from developing countries where the facilities for haemophilia care were scarce. Frank Schnabel was essentially at internationalist. An American by birth, he settled in Canada, and was appointed to the Costa Rican Consulate in Montreal, a position which gave him wide contacts throughout

★ *Frank Schnabel, President of the World Federation of Hemophilia until his recent death, pictured with his wife Marthe during his last visit to the UK, in 1976, when he met the Patron of The Haemophilia Society, HRH The Duchess of Kent. The occasion was the WFH's Third European Regional Congress, held in London.*

the world and enabled him to develop easy communications with groups concerned with haemophilia in other countries.

He was single-minded in his work for the World Federation of Hemophilia, the World Congresses being but the climax of sustained application throughout the year, during which he controlled the extensive administration from the Federation's office in Montreal. We have been singularly fortunate in having Frank Schnabel as founder and then President of the World Federation for, under his leadership, the whole organisation developed effectively as the forum for discussion and action on an international scale.

However, his most valuable contribution had been by his example in living with haemophilia. Many boys and men throughout the world have looked on him as their model as they themselves have learned to cope with haemophilia and rise above the difficulties they experienced as a result of their condition. His example of fortitude, courage and good humour have inspired countless admirers who have been influenced by him. All boys and men with haemophilia who are able to walk through life confidently, with a lift in their step have much for which to thank Frank Schnabel.

At the Memorial Service in Montreal on the Tuesday following his death, the Chairman of our Society was given the opportunity to pay tribute to Frank Schnabel's life and work. It was a moving occasion, for the congregation included representatives from all his interests and those close to him, his business associates, members of the Diplomatic Corps, his friends from Montreal and, above all, his wife Marthe, who had been with him on his international travels and worked as Executive Director of the Federation, and his family who had supported him with such care during his illness.

Now we have come to the end of an era in the life of the World Federation, but the continuing success of its work will be Frank Schnabel's lasting memorial.

MINISTER'S REACTION TO SELECT COMMITTEE REPORT ON AIDS

The Secretary of State for Social Services, Norman Fowler, welcomed the report on AIDS by the House of Commons Social Services Select Committee.

Mr Fowler said: "I welcome the work that the Social Services Committee have done and in particular the focus they have brought to bear on AIDS issues. Their assessment of the position and recommendations will be carefully considered.

"The Government has taken, and continues to take, the threat of AIDS extremely seriously. The Government's evidence to the Committee, including my own evidence to them at Wednesday's session, set out the Government's response. It is summarised below.

"One of the Committee's recommendations is that additional help should be given for the provision of hospice care. I am glad to say that the Government will be providing contributions towards two London projects.

First, the London Lighthouse in West London. £500,000 will be contributed to capital costs and £100,000 to revenue costs for this year.

Second, the Mildmay Mission Hospital in East London. £150,000 will be contributed to capital costs of a new AIDS hospice ward and £50,000 to immediate running costs".

The Government has a four part strategy to meet the challenge of AIDS.

★ Public Health Measures – a range of measures has been taken to make supplies of blood and blood products as safe as possible; and the confidential reporting system monitors the spread of the disease.

★ Public Education – £20 million has been committed to a public education campaign which is internationally recognised as one of the best in the world. This campaign is of great importance as the absence of a vaccine or cure make it a vital weapon against the transmission of HIV infection for the immediate future.

★ Research – the Medical Research Council's request for an extra £14 million has been met in full. This is in addition to the MRC's normal grant-in-aid and the extra £1 million announced last year. It will be spent on a directed programme on a vaccine and on anti-viral drugs.

★ Development of Services – the Government is making sure that services are developed for people with AIDS. All health authorities have drawn up plans at the Government's request. Extra money – £7 million in 1987-88 – has been allocated, in addition to the normal allocations, to help with the special pressures arising in the London area. A further £1.5 million has been provided to help with the immediate costs arising from AZT.

The fifth dimension in the Government's strategy recognises that AIDS is an international problem. The Government believes it is absolutely vital to share our knowledge internationally. There is much to be learnt from the experience of other countries. AIDS is a very serious problem that requires a massive worldwide effort to defeat it. The Government is taking its full part in that battle.

The London Lighthouse project in West London is converting premises in West London to provide 25 hospice beds for people with AIDS, together with counselling and education facilities. The Lighthouse will also provide a base for a Home Support Service. The capital grant of £500,000 is towards the costs of converting and equipping the premises. The revenue grant of £100,000 is towards salaries and administrative overheads.

The London Lighthouse expects to have its premises in use in February 1988.

The £200,000 for the Mildmay Mission Hospital is a contribution towards the costs of a new hospice ward. They will offer nine beds, a counselling service for patients and relatives and a home liaison and counselling service.

CONTINUING OUR LOOK AT . . .

THE WORKING PARTIES

The Case Committee
Presented by Dr L Kuttner

In the last issue of *The Bulletin* we examined the Publications and External Relations working party. This time, I shall try to give you an idea of the work done by the Case Committee, of which I am convenor.

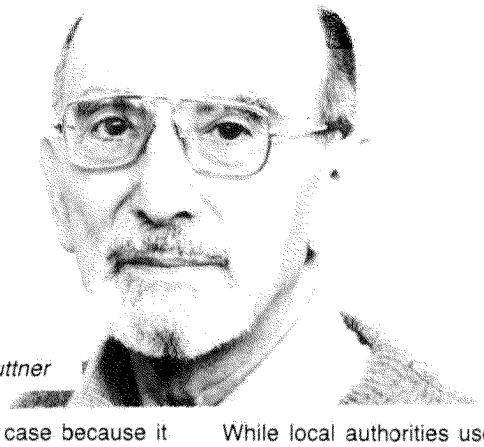
In fact, this committee is about as old as The Haemophilia Society itself and came into existence when the Society was founded, as we were fully aware of the need to deal with cases of hardship and the disadvantaged amongst our own people.

It is perhaps one of the least known, but most important, of our sub-committees. Since the start of the Society, it has been our principle that help to and care of our own people comes long before any other assistance, whether it be research or anything of similar nature. We have tried always to live up this doctrine and over the years this has become a very important function of the Society.

The Case Committee meets monthly and finds that each month many cases are awaiting its decision. The number of cases is growing continually and the amounts of money required are becoming quite a strain on the finances of the Society. What used to be a small demand on our resources a few years' ago is now approaching many, many thousands of pounds annually.

No specific examples can be given (since the Case Committee's work is confidential) and no case is discussed or mentioned outside the Committee's meetings.

Cases come to us from every part of the United Kingdom. They cover the need for telephones, retraining, transport, dietary assistance, domestic help (redecorating, refurnishing, etc.). Most request come from single parents, widows or widowers, unemployed parents and, at times, from old people, all of whom are either people with haemophilia, who have haemophiliac children – or have other blood-disorders, such as V.W. The requests usually reach us via social workers or other official bodies.



★ *Dr. L. Kuttner*

had to refuse a case because it appeared not fully substantiated.

Of all the cases we had to deal with over the last year, the most tragic have been requests for funeral expenses, where parents or wives had not the money to bury their loved-ones who had died of AIDS. All their available money had been spent on their family before the death.

Social workers and others praise the speed with which we make our decisions and provide the cash when it can do most good. The need to help our own people is growing greater daily and the money needed to fulfil this requirement is quite considerable and will increase. For example, at our last Case Committee meeting on 14th April, we required £2,632.00 for 18 cases.

I can say no more about the work done by the Case Committee, but hope that this will give you all some idea of what we do, and that you will help us to do what we feel must be done.

The working parties in general, and the Case Committee in particular, can only function successfully with your generous support.

While local authorities used to be quite generous, some of them give less help now than previously. For instance, some no longer help with the installation of telephones or pay travelling expenses to and from hospitals. Some of the cases brought to us are so heart-breaking and upsetting that they stay in your mind for a long time and often make you wonder whether your decision was right, particularly when you

SOCIETY TIES

NAVY BLUE is the new colour offered in the Society's tie range, which adds to the other two colours in the Gift Catalogue. The navy blue ties were orginally sold by the regional groups some time ago, but have been re-introduced due to popular demand. The order code T.03 remains the same but please state Navy Blue. All the ties are priced at £5.50, plus the normal charge of £1.50 postage and packing.

The Pines, Bishop's Castle, Shropshire

Self-catering holiday accommodation for disabled persons, in groups or families, in attractive, specially adapted house in the incomparable South Shropshire countryside. Indoor and camping accommodation available to December. Swimming, fishing and a wide range of countryside activities available within easy distance. Own transport essential. For particulars apply to: *Mrs. Maureen Jones, The Bungalow, The Pines, Bishop's Castle, Shropshire, SY9 5JY. Telephone Bishop's Castle (0588 638234) or The Secretary, The Pine Trust, The Shirehall, Shrewsbury, SY2 6ND. Telephone Shrewsbury* GRO-C .

PARLIAMENTARY REPORT

On May 15 the Secretary of State for Social Services was asked (1) when his Department first became aware of the threat to haemophiliacs from imported factor VIII in relation to: *(a)* hepatitis and *(b)* HIV virus;

(2) when heat-treatment of imported factor VIII was initiated; and when all haemophiliacs were first able to obtain access to safe factor VIII;

(3) what was the original completion date proposed for the blood products laboratory at Elstree; what is the completion date now; when Britain will be self sufficient in blood products; and what are the reasons for the development slipping behind schedule.

In reply the Secretary of State replied that it became evident in the United Kingdom in 1974 that the use of imported factor VIII was associated with non-A, non-B hepatitis. Progress to eliminate this risk has been limited because the agent responsible has not yet been identified. Clinical trials of factor VIII are in progress to assess if certain heat treatments inactivate the agent.

Evidence emerged in the United States of America in 1982 that haemophiliacs were contracting AIDS and although the mechanism of infection was not known, it was presumed that it had been transmitted through the use of blood products such as factor VIII.

The first account of experimental work which showed that HIV in factor VIII could be inactivated by heat treatment was published in October 1984 ("Morbidity and Mortality Weekly Report 1984", Vol. 33, 589-91). This work led to the National Haemophilia Society of the United States of America advising that heat-treated factor VIII should be considered for haemophiliacs even though its protection against AIDS remained to be proven. Similar advice was published in the United Kingdom in December 1984 (*Lancet* 1984, Vol. 2 1433-1435). However, not until February 1983 (*Lancet* 1985, Vol. 1, 271-272) was evidence published which showed that heat treatment of factor VIII actually reduced transmission of HIV to haemophiliacs.

ONE TO REMEMBER . .

RAW SQUID? 'NO THANKS!' SAYS RICHARD AS ENDURANCE TEST WEEKEND TRIES HIS EVERY STRENGTH

LEICESTER'S GRO-A had to take a couple of days off work after a strenuous activity weekend, around the area of Blackburn in Lancashire, which was to decide if he and five others from his factory would qualify for 'Operation Raleigh'.

The scheme, as you may well know, is run under the auspices of 'The Prince of Wales' Trust' and allows young people from all walks of life to travel abroad to take part in projects destined to help others, mainly in poor countries.

GRO-A who's disabled, works for Remploy in Leicester, and is a keen Society fundraiser in that part of the UK. Sadly, he reports that he didn't pass the selection test but, by all accounts, had a jolly good stab at it – despite one particular exercise which turned his stomach, quite literally. GRO-A had to eat (wait for it) . . . raw squid!

The weekend started at 9 am on a Saturday morning and no-one knew quite what to expect. There were 50 taking part in the exercises, who were split into four groups. Each group had three disabled people and those who could not walk (over three miles of countryside) were carried – and it was the responsibility of the disabled participants like GRO-A to write out the reports of each group achievement.

But the far more demanding tests GRO-A and his fellow disabled friends encountered included crossing a river on a rope and abseiling down a quarry.

Plastic shelter

Overnight shelter consisted of a large, plastic sheet, and food, besides the raw squid, included rice, porridge oats, cloves of garlic and something which resembled a cucumber!

GRO-A takes up the story:

"After putting up the shelter, weighed down with rocks on a dry stone wall, we cooked our meal on the small paraffin stove which we had to be careful with as we only had four matches. We fried the squid, boiled the rice and the vegetables, and flavoured the whole lot with garlic.

The rest of Saturday evening was spent doing logic, memory, sketching tests and one climbing test, using a rope in the gym, and an exercise where you followed a string through an obstacle course, blindfolded. The course apparently had a beach (seemed like tyres to me!) which was prone to tidal waves. 'Oh dear, number six you're a bit slow. Here comes the wave', the tidal wave being a bucket of cold water!

"After being told that that was the end of the day's exercises, we squeezed into our bivouac at about midnight, only to be awakened forty-five minutes later to cries of 'get up quick, there's been an accident!' and a run up a never ending hill to where our *patient* lay, all fourteen stone of him. We then had to carry him back to the centre and give him basic first aid.

Sleep

"Following that, two more tests, one of which was on observation. I must admit to having no idea what colour tie Prince Charles was wearing in his second appearance in the 'Operation Raleigh' video. My mind was on sleep. After all, it was 3 am.

Having been told it definitely was *good night*, ('well *good morning* really') we returned to our shelters, *only to find they had been knocked down and soaked with water!*

"A 'bright and early start' at 6.30 am. Nobody looked bright. At breakfast we did not have time to get the stove going so it was cold porridge oats. It definitely had a squid-like taste from our home-made pans and spoons. Not enough water to wash in, you see.

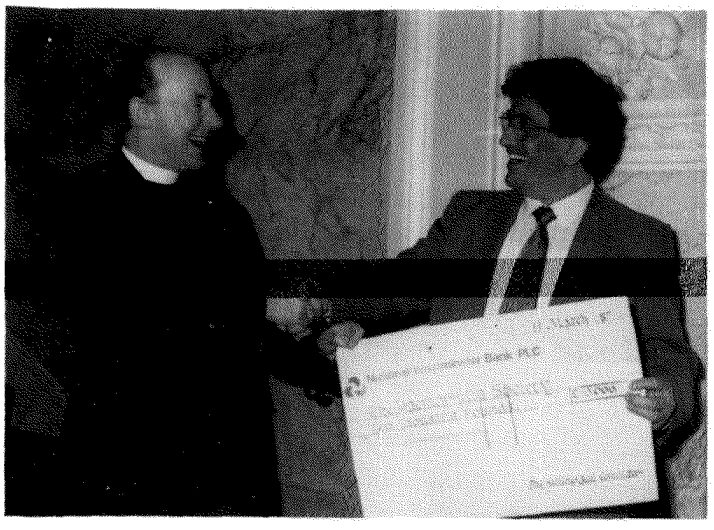
Funny sketch

"After some 'now get out of that' types of tests, involving logs, string, buckets, etc., we were asked to give an individual talk on anything we liked and then each group was asked to prepare something about the weekend and present it to the rest. Every group chose to make a short funny sketch. These must have been fairly funny as even the instructors, who were not noted for their sense of humour, were laughing. After that our numbers were taken off us and we were allowed a swim. Then, up to the dining-room for a hot meal, with orange juice to drink. After that the certificates were handed out."

GRO-A says that more disabled people should go in for 'Operation Raleigh'. It still has two years left to run and even if you do not get on the expedition itself you can learn a lot from the weekend GRO-A certainly did!

★ *Looking none the worse for his experiences* – GRO-A

GRO-A



A BIG, *BIG* CHEQUE!

A hearty *thank you* and handshake from the Society's Chairman, the Revd. Alan Tanner, as he receives a cheque for £10,000 from Brian Bilcliffe, Chairman of the 1986 Airlines Charity Ball Committee.

The proceeds were raised from the glittering social event at the Royal Albert Hall last November, well reported in the previous issue of *The Bulletin*. In addition to this amount, Dave Parker, Freight Manager of Quantas Airlines, raised another £2,429 from a personal sponsored slim.

PLEASE NOTE THAT THIS YEAR THE AIRLINES CHARITY COMMITTEE WILL BE HOLDING A SIMILAR EVENT, AGAIN IN AID OF THE HAEMOPHILIA SOCIETY. THE VENUE, ONCE MORE, IS THE ROYAL ALBERT HALL AND THE DATE FOR YOUR DIARY IS* FRIDAY NOVEMBER 27TH. Further details will follow in due course *(DW).

Mild Haemophilia

by Dr B.T. Colvin

MILD HAEMOPHILIA is the term used to describe the 60-per-cent of patients with X linked factor VIII or IX deficiency who do not, as a rule, bleed "spontaneously" into joints and muscles.

Of course, the "spontaneous" bleeding of severe haemophilia involving these moving parts is probably the result of minor injury to blood vessels in most cases. But whatever we may think about the finer points of the argument, the bleeding certainly feels spontaneous to the severely affected patient.

Not much factor VIII or IX is needed to avoid the worst effects of the deficiency and anyone with more that two or three per cent of the average normal level (2-3 i.u./dl) is likely to fall into the mild category. At the other end of the spectrum are the very mildly affected patients with factor levels up to about 40 per cent (40 iu/dl) and it is important to remember that many carriers of haemophilia are subject to abnormal bleeding if their factor levels lie below the normal range.

Once the bleeding has started it may then continue indefinitely, even in mild haemophilia, until something effective is done. Mild haemophilia does not mean mild bleeding. Similar problems are also seen in other clotting factor deficiencies, of which factor XI deficiency is perhaps the commonest, but platelet disorders usually behave rather differently. *Von Willebrand's Disease* shares features in common with mild haemophilia and the platelet disorders, but is not the subject of this article.

If there is a strong family history of haemophila the diagnosis is often made at birth, whatever the severity, but mild haemophilia can be diagnosed at any age. In childhood, circumcision if the most immediate hazard but the sharp toy in the mouth, the fall onto the face when the teeth can damage the lips and the dramatic effects of tonsillectomy are all familiar to haematologists. Accusation of non-accidental injury (child battering) is very upsetting for mothers and is not entirely confined to severely affected families.

As children get older they begin to take part in organised sport and this can precipitate bleeding. A stretching injury to the muscles at the back of the lower abdomen (usually the *psoas muscle*) can be very difficult to diagnose and is easily confused with appendicitis if the pain is on the right hand side. A single intramuscular injection of vaccine or antibiotic can also cause serious damage during dental extraction and is a more obvious and traditional enemy of the patient with haemophilia.

As adults get older they become more and more likely to need an operation of some kind and the oldest previously undiagnosed patient I have seen was a man in his late 60's who had nearly died after a prostate operation. His grandson, who had never had abnormal bleeding of any kind, was also found to be affected.

Once a significant bleed has occurred the correct diagnosis is not always made. The family of a patient may think of themselves as "bleeders" but fail to associate their bleeding tendency with a real disorder. Where the symptoms of haemorrhage are confined to pain and swelling, as in the muscle bleeds mentioned above, the diagnosis is often unclear and there is a grave risk that exploratory surgery may be undertaken with potentially disastrous results.

Few doctors understand that months may separate the injury itself from the decision to seek medical advice. Thus one elderly man under my care became gradually unconscious three months after a minor head injury and was found to have a blood clot on the brain. A few years later his grandson twisted his ankle at a Christmas disco and watched the badly swollen joint get gradually worse until he eventually came for advice at Easter. Both made a full recovery with appropriate treatment.

Once the suspicion of haemophilia is raised the results of laboratory tests need to be interpreted carefully. It is well known that the factor VIII level rises as a result of stress and in very mild haemophilia the value may be almost normal when measured at the time of active bleeding. This means that in doubtful cases careful follow up is necessary with reassay of clotting factors when the patient is well again.

★ *Dr Brian Colvin.*

If haemophilia is diagnosed in childhood then my policy is to offer all children regular follow-up in our combined paediatric clinic, whatever the severity of the disorder. This enables us to give the necessary immunisations, including hepatitis B vaccine, subcutaneously, and to give advice on treatment, dental care and schooling from the earliest age. The clinics also establish a relationship between the hospital and the family which can be invaluable if the child falls ill or requires surgery at any time. As the boy develops his parents can be given sensible advice about what he should be able to manage at school.

Boxing and rugby football are really too hazardous for any child with haemophilia but for other sports it is best to see how things go, allowing the patient to find out for himself the things he enjoys and is capable of. If soccer and cricket prove too much then running, swimming and tennis can be encouraged without the young person feeling he has been deprived of opportunities by his parents or doctors. All sorts of problems turn up which, while not strictly related to haemophilia, are best dealt with by the team approach in which paediatricians, dentists, nursing staff and social workers play a key role. For mildly affected children a six-monthly appointment may be quite sufficient and is well worthwhile.

For adults it is traditional to offer yearly follow-up to the less severely affected patients, but the success of this approach depends very much on the nature of the patient, the distance from his home to the nearest haemophilia centre and the type of work he does. The patient who fails to attend often conveniently forgets about his haemophilia and may ignore a serious bleed or fail to interpret it correctly. He eventually comes to the hospital in a worse condition than would a more severely affected patient who is used to the effects of his disease. It is also not uncommon to see a patient bleeding from a dental socket having had an extraction without telling the local dentist of his haemophilia because he hoped "it would be all right". Other potential dental errors include the use of inferior dental blocks (injections of local anaesthetic around the nerve) and the prescription of aspirin for pain.

When seeking employment the patient with mild haemophilia is much more likely to conceal his condition from his employer and this is, perhaps, understandable when one considers how difficult jobs are to find these days. It is true that once established in a post with a good work record a sympathetic employer may be more inclined to take a lenient view of absence from work caused by a serious bleed or the need for surgery. It must be understood, however, that failure to disclose the illness could be regarded as grounds for dismissal and that no doctor can really advise non-disclosure as a sensible course.

It is particularly difficult to advise a young man whose heart is set on joining the Police, the Fire Brigade or the Armed Forces when it is clear that, despite the mildness of his condition, he is really unsuitable for the work and is running a serious risk by applying without disclosing his haemophilia. One can only argue strongly against this course of action while explaining that the haemophilia centre director will not, and indeed cannot, inform anyone of a patient's condition without permission. It is equally important to make sure that patient knows he can still return for help and treatment despite the fact that he has ignored his doctor's advice.

Very similar issues are raised by the need to take out insurance policies. The fact that someone has mild haemophilia does not alter the necessity of filling out the proposal form fully and correctly and failure to do so can result in the policy issued being declared void. The consequences of this happening to a patient who has taken out life insurance, mortgage protection or holiday insurance for trips abroad can be catastrophic. Fortunately, the Haemophilia Society is able to give excellent advice in this field but for those who do decide to organise their own cover it is essential to read the proposal form very carefully. For instance some holiday policies ask only that a person does not travel against his doctor's advice while for others it is necessary to certify either that there is not illness and that no medication is being taken or else declare the problem. Haemophilia centre directors may have to advise the insurance company for the second, less attractive, type of policy so it is worth making plans well before the trip begins.

Mild Haemophilia
(continued)

The patient with mild haemophilia does not often require treatment. Joint damage is unusual and inhibitors are rarely a problem. Home treatment is only occasionally worthwhile for those who enjoy very vigorous sports, bleed a little more frequently than average or live deep in the countryside. Until the early 1970s most patients were treated with cryoprecipitate and many centres continues to use this material for mild haemophilia even when freeze-dried concentrates became available. After 1977 vasopressin (DDAVP) was introduced to stimulate the body's own factor VIII level in patients with mild haemophilia A, carriers of haemophilia A and *Von Willebrand's Disease*.

The great advantage of this approach was that blood products could be avoided altogether in many situations, thus removing the risk of hepatitis or other virus infections. DDAVP is given with tranexamic acid, which prevents the breakdown of blood clots and the combination is ideally suited to minor surgical procedures such as dental extraction but can be used for tonsillectomy, hernia repair and other operations. Unfortunately, the effect of DDAVP only lasts a day or two before the body's factor VIII stores are exhausted and this means that for bad injuries or major surgery factor VIII replacement therapy still has to be used, especially for those with lower factor VIII levels.

Consequently a number of patients with mild haemophilia have developed acute hepatitis and anyone who received freeze-dried concentrates before 1984/85 is bound to have shown some signs of liver infection, even if this caused no symptoms at the time. Some patients have also developed antibodies to the human immuno-deficiency virus (HIV) and are thus at risk of developing AIDS. From this point of view all patients with haemophilia have run the same risks but it is understandable if mildly affected patients feel particularly bitter and wonder whether their concentrate treatment was really necessary. Fortunately, now that the newer NHS and commercial concentrates are available, the risk of virus infection has been much reduced and possibly eliminated so that this form of treatment is probably safer than cryoprecipitate. In the future synthetic factor VIII is likely to add an extra degree of safety.

Another aspect of haemophilia care that has advanced very rapidly in recent years is genetic counselling and antenatal diagnosis. No-one can alter the fact that a third of all babies with haemophilia are born to parents without a family history but, for those who know of the risk, accurate advice and reliable prevention is now usually available. Initially it was only possible to measure factor VIII or IX levels in the blood of women who might be carriers and to take blood from the unborn child in the middle of pregnancy for antenatal diagnosis.

More recently, the development of gene probe techniques has given greater certainty to carrier detection and allowed antenatal diagnosis after ten weeks of pregnancy for many families. As haemophilia has the same severity from generation to generation it is possible to reassure mildly affected families that any children born with haemophilia will have the same type of problems as existing affected relatives. It is then necessary to discuss with prospective parents how much information they need and how far they are prepared to go in preventing the birth of an affected child.

Decisions can be made on the selection of appropriate investigations, bearing in mind that for gene probe techniques to be successful many members of the family may need to attend for blood to be taken and that this approach takes a great deal of laboratory time and effort. All antenatal diagnostic methods carry some risk to the unborn child and are clearly not appropriate if there is not intention to terminate an affected pregnancy. It is therefore wise to consult the haemophilia centre director as soon as advice on carrier detection is needed so that sensible decisions can be made, unaffected by emotion or urgency. It is not the function of the centre to impose decisions on the family but to help women and parents to achieve the result they desire after everything has been explained to them.

Haemophilia is never easy to live with. Those of us who care for patients must remember that our efforts on behalf of the most severely affected patients should not blind us to the needs of the others. When patients with mild haemophilia need treatment they require all our care and attention. When decisions have to be made they are just as difficult as for any of our other patients, even if their nature is a little different.

PERSONALITY PROFILES

GRO-A

GRO-A has been a member of the Haemophilia Society for 35 years and is a member of the World Federation of Haemophilia.

He joined Sheffield and District Group as a committee member and then served as Chairman for two years before being elected to the Executive Committee in 1985.

GRO-A at 41, is a member of the Case Committee and the Treatment and Care Working Party and has just recently taken on the demanding role of Group Liaison Officer.

He therefore takes a keen interest in Group activity all over the UK yet still finds time to serve as a committee member of the Sheffield Group.

He was born in Cleethorpes at the end of the war and, in keeping with most haemophiliacs of that era, suffered a much-interrupted education owing to the usual bleeding problems which could not be treated. GRO-A attended the Lord Mayor Treloar College, gaining GCEs and a City and Guids diploma in Radio and TV Servicing. He went on to study electronics at colleges in Grimsby and Lincoln, eventually attaining a full Tech. Cert. in Electronics.

Today, he is the manager of a Service Department which specialises in the bulk repair and maintainance of electronic appliances for several major third-party contracts.

GRO-A has two brothers (one of whom also has haemophilia) and is married with two children, aged nine and seven. His hobbies are, he says, too numerous to mention in total but top of the list are photography, music (he played drums for a number of years on the northern clubs circuit), astronomy, gardening, and the history and art of magic. It perhaps follows, therefore, that this very active man is a member of the Northern Magic Circle and The Supreme Magic Club of Britain.

Finally, GRO-A is also an associate member of the Society of Electronic and Radio Technicians.

GRO-A

GRO-A

Londoner GRO-A is 31 and as we went to press was battling as SDP candidate in Hackney, London, GRO-A. He works as a Public Relations consultant which, in practice, means that he advises companies on the workings of Government and the EEC.

Like many other members of the Executive Committee GRO-A who has severe haemophilia, went to the Lord Mayor Treloar College until he was 16 – and then to St. Edward's School, Oxford. Simon passed out of Durham University with an honours degree in Economics and Politics.

Within the Society he has been chiefly working on its responses to the AIDS crisis, advising on relations with the press and the Government. In what spare time his job and the Society leaves, Simon is busy canvassing.

GRO-A

LOOKING BACK OVER 37 YEARS OF HAEMOPHILIA CARE IN THE UK

A REPORT ON THE 1987 HAEMOPHILIA SOCIETY LECTURE GIVEN BY DR CHARLES RIZZA MD, FRCP.

★ *Dr Rizza takes to the lecture rostrum.*

NEWCASTLE'S Crest Hotel was the venue for this year's annual lecture on 28th March which was given by Dr Charles Rizza MD FRCP, who is Director of the Oxford Haemophilia Centre.

Dr Rizza's delivery, which was part of the Seminar Weekend, took a long and detailed look at the way haemophilia care in Britain has developed since the start of the 1950's and was appropriately titled THE HISTORY AND ORGANISATION OF HAEMOPHILIA CARE IN THE UNITED KINGDOM. It also provided a personal view on how he saw matters evolving in the years to come.

For those unable to attend, we present his paper here . . .

"EARLY IN 1950 Sir Weldon Dalrymple Champneys, who was Deputy Chief Medical Officer at the Ministry of Health, and was later to become president of your Society, wrote to Dr. Macfarlane at Oxford and Dr. Wilkinson at Manchester expressing his concern at the problems experienced by haemophiliacs and suggesting a meeting at the Ministry of Health to look into the matter.

This move was prompted in part by Sir Weldon's interest in the welfare of haemophiliacs and in part by approaches made to him in the late 1940s by several haemophiliacs who had formed themselves into an International Association of Haemophiliacs. I presume that that Group was the forerunner of your Haemophilia Society. Three weeks later on the 13th March, 1950, a small group met and it was agreed that with resources available from the newly set up National Health Service it ought to be possible to do something on an organized basis for patients with haemophilia. It was suggested that such arrangements might come under three headings:

- **Advice to the patient on how best to look after himself and avoid injury.**
- **Suggestions to the medical profession about treatment and supervision,** and
- **Education of the general public on the need for special consideration for haemophiliacs.**

There was unanimous agreement amongst those present that with very few exceptions medical practitioners did not know how to handle haemophilia.

FIRST STEP

As a first step it was agreed that there was a need to establish in each region one or more Centres (depending on the size of the region) to which patients could be referred for diagnosis, treatment and advice. A number of (3 or 4) Centres was suggested in the first instance with the numbers increasing to 10 or more according to need. The plan initially was to find doctors who had knowledge, interest, resources and enthusiasm and then to induce the regions to provide facilities. It was recognised that once a scheme was going, important activities would need to be organised, in particular the hospital almoners would need to be involved, transport for patients arranged, and labour exchanges would need to collaborate as would the education authorities.

A more formal meeting to discuss the social and medical problems of haemophiliacs was held at the Ministry of Health in October 1950. This was attended by 24 individuals and included representatives from the Ministry of Health, the Ministry of Education, the Medical Research Council and doctors who had haemophiliacs under the care. The meeting agreed that recognition of Centres was desirable and all of those present agreed to make facilities available for care of haemophiliacs. In this way the population of London, Newcastle, Sheffield, Oxford, Birmingham and Manchester was covered.

LIKELY CENTRES

Cambridge, Cardiff and Bristol were also proposed as likely Centres. It was further agreed that those doctors concerned should meet regularly to exchange information. In addition to recognising the need for a number of special Centres up and down the country to treat haemophiliacs, the Committee agreed that haemophiliacs should be issued with a special card and that information should be disseminated to local health, education and welfare authorities and labour exchanges explaining the haemophiliacs' problems and how they should be managed. It was also suggested that a pamphlet outlining the problems of the haemophiliac should be drawn up by medical experts and sent out to the medical profession.

At about the same time as those meetings were going on at the Ministry of Health, the Medical Research Council in 1951 appointed a small and informal Working Party to advise on the problems of haemophilia and in 1953 this group was reorganised as an official committee of the Council. The Committee chaired by Dr. Wilkinson of Manchester comprised those who had attended the M.O.H. meetings. It reaffirmed what had been proposed at the earlier Ministry of Health meetings and suggested that a number of clinics and laboratories throughout the country should be invited to act as Reference Centres with the object ot ensuring uniformity of diagnostic standards and co-ordinating exchange of information. It was also agreed to keep a register of haemophiliacs which would prove helpful to patients and research workers alike. By 1955 the list of Centres had grown to 19.

REGULAR MEETINGS

The MRC committee met regularly to discuss advances in haemophilia research and through its AHG Sub-Committee encouraged collaboration and research into development of human and animal AHG and into development of assay methods. This was a very important and fruitful time in our understanding of haemophilia and in the development of treatment. Calculations were made of the number of haemophiliacs in the country and for the first time thought was being given to the best way of managing different kinds of haemorrhages. In particular the importance of treating haemarthroses was becoming apparent and plasma was being used.

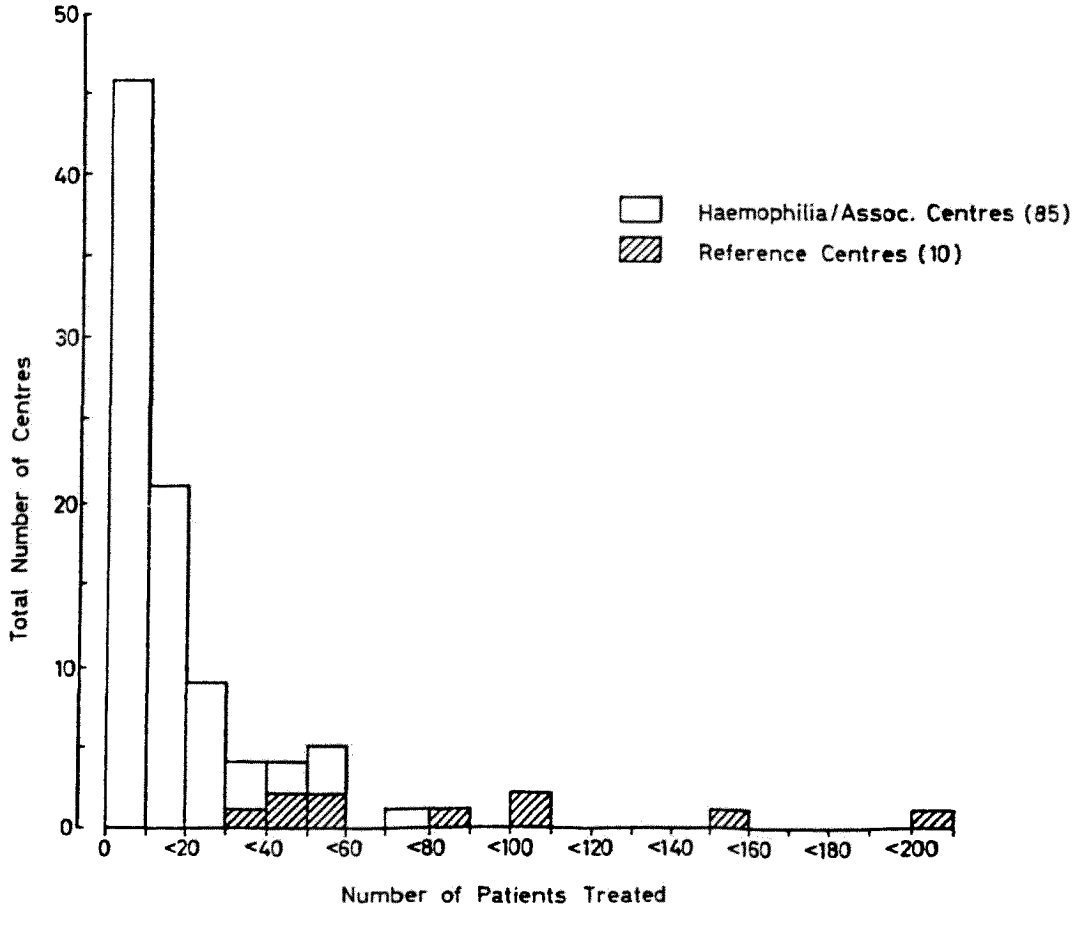
Before this time work in haemophilia was mainly concerned with basic research into the condition, was laboratory orientated and had little to offer in the way of effective treatment. The service offered was diagnostic and supportive and this was provided mainly by laboratory haematologists who often did not have access to beds for treating their patients. This caused some problem later when therapeutic concentrates became available in larger amounts and Centres often found themselves without beds and without staff trained to treat patients.

In 1964 the responsibility for overseeing the organisation of haemophilia care passed to the Ministry of Health and in 1966 the M.R.C. Haemophilia Committee was disbanded. At about the same time the prospects of effective treatment for haemophiliacs was greatly improved by the introduction of cryoprecipitate and the Haemophilia Centres which had been previously concerned mainly with carrying out diagnostic tests and registering patients were now able to provide a treatment service at short notice and to carry out major surgery safely.

As a result of these major advances in treatment it was necessary to review the organization of haemophilia care and to define the function of Haemophilia Centres. In 1968 the Ministry of Health produced its health memorandum HM68(8) concerning "Arrangements for the care of persons suffering from Haemophilia and related diseases." This memorandum contained a list of 36 Haemophilia Diagnostic and Registration Centres that would be prepared to take responsibility for the care of those suffering from haemophilia and related diseases in addition to the essential function of diagnosis, registration and the issue of haemophilia cards. According to the memorandum Centres were required to give a laboratory service and carry out the tests as a result of which exact diagnosis could be made and a haemophilia card issued; provide a clinical service; provide treatment at short notice and provide an advisory service on a wide range of topics of concern to the patients. The service was to include advice on the care of small children with coagulation disorders and on the most suitable education available. The service was also to provide general practitioners who had haemophilic patients on their lists with advice about emergency treatment and the procedure to secure patients' immediate admission to hospital when required.

S.T.C.'s

Three of the 36 Centres, in addition to performing the functions of Haemophilia Diagnostic and Registration Centres were designated as Special Treatment Centres to undertake major surgical treatment of patients with coagulation disorders and were to be available for consultation about all dangerous lesions in those patients. The Centres designated as Special Treatment Centres were at Oxford, Manchester and Sheffield. It was recommended in the memorandum that close contact should be developed between the staff of the Diagnostic and Registration Centres and the appropriate Special Treatment Centre so that the greater experience and resources of the latter could be of benefit to any patient who needed it particularly when planned or emergency surgery was necessary. Finally it was suggested that close relationships should be developed with the appropriate Regional Centre of the Blood Transfusion Service from which fresh frozen plasma would have to be obtained.



In addition to the publication of the memorandum HM68(8), 1968 saw the first meeting of the U.K. Haemophilia Centres' Directors' Organization as we now know it. This was held in Oxford on 1st October 1968 to mark the opening of the Oxford Haemophilia Centre's new buildings and was attended by 36 doctors. Those included Haemophilia Centre Directors, Blood Transfusion Directors, plasma fractionation workers and a representative from the Ministry of Health.

The Haemophilia Society did not attend the first meeting but has sent representatives from 1974 onwards. Dr. E.K. Blackburn was elected as Chairman of the Group. Items discussed at that first meeting included: *Design of a new haemophilia card; what type of patient should receive a haemophilia card; record keeping at Haemophilia Centres and last but by no means least, the classification of Haemophilia Centres.*

The minutes say that "the discussion on this topic was inconclusive".

ANNUAL MEETINGS

The Directors thereafter met yearly and I think it is fair to say that there have been few occasions on which organization of haemophilia care has not appeared on the Agenda. Clearly there was a feeling that the organization of care was still not good enough. At about the same time as the 1968 meeting of Directors in Oxford representatives of the Haemophilia Society approached Professor Dacie in London to say that they were unhappy with the arrangements for haemophilia care in London and that they would rather see a small number of high quality well staffed Centres than the large number (12 in fact) of Centres of varying quality designated in HM 68(8). A meeting was held some time later at the Department of Health to try to rationalize the number of Centres in London.

This was attended by all the London Directors and consideration was given to a 3-tier system of haemophilia care for London. It was suggested that there should be:

(a) Diagnostic Centres which would diagnose the disorder and issue cards but would not attempt to give a 24-hour clinical service.

(b) Diagnostic Treatment Centres which would undertake all the functions of Diagnostic Centres but also would provide a clinical service

(c) Special Treatment Centres. As there were no Special Treatment Centres designated in London

under HM 68(8) it was recommended that the Royal Free, St. Thomas' Hospital and Great Ormond Street should be Special Treatment Centres.

That particular three-tier system found no favour with the London Directors but the idea of a 3-tier system remained in the air and in 1974 a draft document on reorganization of Haemophilia Centres throughout the country was discussed by the Haemophilia Centre Directors. This had been drawn up following consultation with an Expert Group on Haemophilia and proposed a three-tier system of Haemophilia Centres. The document was discussed in detail in November 1974, put to vote and accepted by all except three of the Directors. This draft document went on to become HC 76(4) which is the current momrandum relating to organization of haemophilia care in this country. The memorandum set out revised arrangements for care of haemophilia and related disorders based on a three-tier system revised the criteria for designation of Haemophilia Centres and asked Regional Health Authorities to review Centres at which treatment was available to patients.

FUNCTIONS DEFINED

The functions of Haemophilia Centres were defined as were those of Reference Centres. To avoid inconveniencing patients, Centres which had been designated in 1968 but which could not fully meet the new criteria, would continue to be recognized for giving emergency treatment and would be known as Associate Centres. These Centres were to be linked with a convenient larger Haemophilia Centre and together offer patients a full diagnostic treatment and advisory service to haemophiliacs and their families. The number of Centres listed in the appendix of HC76(4) was 74, seven of which were Reference Centres. Ten years later, there are now 109 Centres, of which 10 are Reference Centres.

The Centres vary considerably in size, staff and number of patients treated. Except in a very few instances Haemophilia Centres are part of or closely linked to Departments of Haematology and the directors are haematologists, usually having a special interest in haemophilia. In general the Reference Centres are larger than the other Centres although there are two or three non-Reference Centres which deal with more patients than some Reference Centres. The table on the previous page shows the number and distribution of severely affected patients treated at Centres in the U.K. during 1985. The most striking feature is the large number of Centres who see only a few patients in any one year.

Indeed, five Centres are reported to treat no severely affected patients and another 46 treat fewer than 10 severely affected patients in a year. It has been suggested, in line with W.F.H. policy, that any Centre which treats fewer than 10 severely affected patients cannot have the expertise, organization and staff to offer a satisfactory haemophilia service and therefore should not be listed as a Treatment Centre. This may be the case sometimes. On the other hand many small Centres given an excellent very personal service thanks to the special interest of the director and his colleagues.

As you well know, it is because of considerations such as this that thought is being give to reorganisation of haemophilia care in the country. Before proceeding to dismantle the present system we must ask ourselves what we are aiming to achieve. The answer to that question is, in my opinion, easy. We should be trying to provide all haemophiliacs with prompt emergency treatment and follow-up wherever it is required; provide facilities for safe surgery; provide support, advice and counselling on a wide range of topics including education, employment, genetics, HIV etc. All of those services will require laboratory facilities to enable assays to be carried out for diagnostic purposes or to monitor factor replacement during surgery. Ideally these services should be available as close as possible to the patient's home and it is here that problems begin to arise.

LONG DISTANCES

Many haemophiliacs live at a considerable distance from a large Haemophilia Centre and find it very inconvenient to travel to the large Centres for treatment of day to day bleeds. They prefer therefore to attend the local small Centre for treatment. The problem is slightly less for those patients who are on home treatment although even here there is a danger that the patient will be seen only infrequently by either the small local Centre or the larger Centre. This difficulty can be particularly marked in young haemophilic children as they are less likely to be on home therapy and their parents (usually mother) may find it very difficult to travel long distances for treatment. In such situations a good local Centre is essential if the frequent minor bleeds which are the bane of haemophiliacs are to be dealt with effectively. The problem is how to provide good emergency treatment at the small Centre and at the same time have the facilities and expertise of the larger centre available for treatment of major bleeds, general follow-up, counselling, etc.

If we decide that Centres which treat less than 10 severely affected patients should not be Haemophilia Centres we shall do away, at least in name, with 50 Centres, nearly half of the Centres in the country. As a consequence 200-300 patients, many of whom had received good treatment at their local Centre, will find themselves travelling long distances for treatment at great inconvenience and possibly risk to themselves.

On the other hand, if hospitals which have ceased to be listed as Haemophilia Centres continue to treat patients, and this may well happen, we shall be in a situation where a significant number of patients will be treated outside the Haemophilia Centre Directors' Group. Those doctors concerned will presumably no longer attend the Directors' Meetings, will not contribute statistical information to the group and will not benefit from the education and exchange of ideas that take place at those meetings. In my opinion this would be most undesirable.

At the other end of the scale the role of Reference Centre has been questioned. As you will remember, Reference Centres were originally designated on a supraregional basis and very broadly their function was to provide a specialist clinical and laboratory consultant service for those Centres which wished to use such a service, to co-ordinate educational and research programmes and to co-ordinate statistics. There is a feeling now that haemophilia should be organised

★ *The Revd. Alan Tanner pictured (second, left) at the start of the civic reception with the Lord Mayor and Mayoress of Newcastle, Cllr. and Mrs Peter Laing, with his mace bearer and, on the far right, Dr and Mrs L. Kuttner.*

★ *A reception dinner was held at the start of the seminar weekend in Newcastle, hosted by the Lord Mayor, Cllr. Peter Laing (centre). Pictured sharing an after dinner joke with him is the Society's Chairman, The Revd. Alan Tanner (left) and local haemophilia specialist, Dr Peter Jones.*

on a regional basis that each region should have a Regional Centre which would have responsibility for its Health Region only and not a supraregion.

Haemophilia Centre Directors and representatives of the Society discussed this matter at a meeting last year and it was overwhelmingly agreed that **haemophilia care should be organised on a regional basis and** that each region should have a Regional Centre which would be responsible for organizing haemophilic care, in collaboration with other smaller Centres in the region. There are several consequences of this policy of reorganization if it goes ahead. For example in some regions there may be two hospitals serving as Haemophilia Centres – which one will become the Regional Centre? How will this be decided and what will be the financial implication if one is chosen instead of the other?

In some regions there is no large Haemophilia Centre but a scattering of many small Centres. Which Centre should be designated as the Regional Centre? Does it have the necessary facilities and expertise? If not, would extra funding if it were available make a difference? I suspect not. Expertise in haemophilia care is built up over many years and comes from looking after many patients in a wide range of clinical situations. Moreover expertise must be present not only in the Centre Director but the nurses, surgical colleagues, laboratory staff, physiotherapists, social workers and others involved in patient care.

★ *At a special ceremony during the seminar weekend, local haemotologist, Dr Peter Jones, was presented with a 'thank you' gift from Society Chairman, the Revd. Alan Tanner – a mobile telephone.*

PERSONAL VIEW

My personal view is that every attempt should be made to make the present three-tier system work. It has the capacity for allowing patients to receive their day to day treatment locally and at the same time affords access to more specialized testing, treatment or counselling at large Centres. If the present system or organization has failed, as some suggest, this has been because the guidelines laid down in HM 76(4) have not been observed as closely as they should have been.

This might be largely put right by encouraging the small Centres to liaise more closely with the large Centres and also by encouraging the patients, with backing from the Society, to seek advice from the larger Centres if they are meeting problems at the small local Centre. Probably this should be tried before any attempt is made at wholesale reorganization.

Our primary aim must be to give the patient and his family the best possible treatment and support. That should be our starting point when thinking about any reorganisation and reorganisation should not be seen as an end in itself. Regrettably as with many human endeavours fairly simple objectives and aims have become obscured by other considerations which are not related directly to patient care."

KONTACT KORNER

GRO-A is a 16-year-old haemophiliac who lives in Beauvais (just 70 km from Paris) who would love to do an exchange holiday with someone of about his own age in this country. The teenager has been studying English at school for the past three years.

He has a brother and sister in their 20s, and GRO-A is very fond of drawing, ski-ing and swimming. This young man takes his school holidays in July and August and would like to stay with a family over here for two weeks and then return with his counterpart for a fortnight spent at his home.

GRO-A lives in Paris and is 13. He's been to England (London) once before and is very keen to return to the UK, round about August this year, to stay as a paying guest if necessary, with an English family and would prefer to be in the London area or the Home Counties, close to either Heathrow or Gatwick. GRO-A hopes thereby to improve his English and learn more about our way of life.

GRO-A's mother describes him as a "major degree haemophiliac" but he keeps himself fit and receives treatment regularly from a hospital in Paris.

★ *If you're interested in accommodating either of these two lads this summer please do get in touch with General Secretary David Watters who will supply contact details.*

'Young Disabled On Holiday'

Still with thoughts of summer and getting away from it all in mind, the above is the title of a new charity group which organises active holidays at home and abroad for physically handicapped young people, aged 16 to 30.

The group's Secretary, Rosemary Girdlestone, explains: "We have run holidays in various locations, including Edinburgh, Lancaster, York, Southampton and Wales in the UK as well as in Germany, Spain, France, Ireland, Malta, Lanzarote, amongst others.

Helper

"We use a variety of accommodation – tents, boats, universities, chalets, hotels and so on. All holidaymakers have a helper to suit their needs and that help can vary from tying a shoelace to assistance with washing, dressing, lifting and other personal things".

Holiday Planning

IT'S HOLIDAY TIME again and the Royal Association for Disability and Rehabilitation (RADAR) has recently published some new guides for travellers – for breaks taken at both home and abroad.

'Holidays For Disabled People' costs £2 and is available from branches of W.H. Smith or any good bookseller. It's crammed with information on hotels, apartments, nursing homes and holiday centres, whilst also including sections on activity holidays for the young, transportation and group accommodation.

Priced at £1.50, *'Holidays and Travel Abroad – A Guide For Disabled People'*, is only obtainable from RADAR. The cost includes p & p and the address to write to is: *RADAR, 25 Mortimer Street, London W1N 8AB.* As you might expect, this booklet gives information on travel by sea and air as well as coach services. Contact names and addresses are as widespread and varied as Barbados and Yugoslavia, including those for wheelchair hire and repair.

A third helpful booklet is *'Access at the Channel Ports'*. This aims to take the uncertainty out of routing your journey through the ports on both sides of the Channel and will help you to avoid long walks or steps, and to locate suitable toilets. Details include access to terminal buildings, wheelchair accessible toilets, accessibility of cabins and boarding arrangements, as well as a comprehensive contact directory. The publication, costing £2.50 and again available from the above address, was prepared by a group of old students of the Hephaistos School, Reading, and St. Paul's School in London.

As Rosemary also points out, a holiday away from home can also mean a break for parents or loved ones in the family who would normally take care of the disabled young person. For more information about YDH you can contact Rosemary at GRO-C GRO-C GRO-C

★ **LEWISHAM GROUP are looking for items for their bric-a-brac stall and prizes for their tombola at Blackheath Fayre in early June. Any donations, please, to the Secretary, Heather Hodgkinson, at GRO-C**

A MOTHER'S STORY

Coping with the problems of children with haemophilia

BRINGING UP a child suffering from haemophilia has its problems for any parent. They are always on their guard to ensure that the youngster does not put him or herself in a position where a bump, fall or collision might spell danger.

In their own words, three mums whose youngsters attend a children's clinic at a major hospital in the UK recount their own experiences and how they have come, or are coming, to terms with the blood disorder in their offspring.

They each had to start from scratch in the learning and caring process. None of their children is HIV positive and so AIDS does not crop up in these warmly written pieces. However, we hope the three stories which follow offer encouragement and comfort to other parents.

GRO-A

Haemophilia is something very new to me. My son, who is eighteen months old, was diagnosed a severe haemophiliac at nine months and since then it has not been easy for myself or my family.

As GRO-A is a severe haemophiliac, we have had many trips to our local hospital and the children's clinic. The care and attention GRO-A receives locally, especially from the nursing staff, cannot be faulted. We have had some difficult times and some very frightening times which I hope are all behind us. I now believe that all the difficulties we had were simply a lack of understanding of haemophilia on my part and also by the young trainee paediatric doctors. We now have a very good rapport with the Haematology Consultant and the paediatric team and the treatment GRO-A now receives is very good.

Our clinic appointments have been of tremendous benefit to me. The Haematology Consultant and his supportive staff have helped me over the last nine months with their willingness and patience, explaining to me and my family about haemophilia. The Sister has been a life-line. She has helped me tremendously and the knowledge that she is there and can be contacted if I have any problems is very reassuring.

The Sister and the Social Worker have helped to form and co-ordinate the parents' group. This group meets three times a year, and at these gatherings we feel at ease to discuss any problems or difficulties we are experiencing, and it is nice to know that perhaps other parents have experienced the same problems and that things have worked out. It is a great help, I feel, to all those who attend.

PLACID AND GENTLE

I am very fortunate in that Jamie is a very placid and gentle little boy – totally different in character from his older brother GRO-A my elder son, although being five, copes well with the pressure of having a younger brother who is very special. There are times when, like all brothers, they have a set-to over a book or toy. I try to stand back and let them sort it out, as I feel it is very important not to be over-protective. GRO-A goes to toddler groups and crêches, and he is learning to stand up for himself and to be as normal a little boy as possible.

GRO-A

As a family, we visit the Children's Clinic three to four times a year. When we arrive our son, GRO-A, is seen by the Clinic Sister, who will then record his weight and height. We then sit in the waiting room decorated with 'Winnie the Pooh' cartoons while the children play in the toy room. This gives us the chance to chat to other families in the same situation as we are.

In the consulting room is the team of people, headed by the Consultant Haematologist, who will be involved in GRO-A s treatment at some time. Because GRO-A has been able to meet everyone involved, he is quite happy when it comes to visiting the Dental Institute for his dental care, or receiving visits from the Haemophilia Sister or Social Worker, and when an in-patient on the children's ward there is usually a familiar face around. GRO-A is then examined by the Consultant Haematologist and the Consultant Paediatrician. Questions are asked about treatments, hospital visits and any illnesses or problems since the last clinic. We then have the chance to ask any questions of our own and also to arrange for situations outside the hospital, such as starting school and any letters that may have to be written and follow-up visits to be arranged.

PARENTS' GROUP

Out of the clinic came a parents' group, which is held in various parents' homes. With the Haemophilia Sister and Social Worker from the clinic, families in the same situation started to meet about four times a year to discuss and share their problems. The two consultants came to one of our meetings to understand what they were about and how we all felt about the hospital and treatment received, and if there was anything we would like changed.

We started to learn about *'Home Treatment'* through the clinic. It was discussed at one of our earlier clinics whether we would be happy to treat GRO-A ourselves at home. After that it was talked about at nearly every clinic, until we all felt that the time was ready GRO-A was just four and a half when the Haemophilia Sister visited on a weekly basis to teach us the procedure of home treatment. At first we practised on each other and willing members of our family, until gradually introducing it to GRO-A With back up from the Sister with frequent visits, arrangements of supplies and equipment and the encouragement of the Clinic Team, GRO-A is now having full home treatment at the age of five.

This account is based on our experience of the 'Children's Clinic' and the outside help attached to it. Five years ago we knew next to nothing about haemophilia, but now we feel competent and confident in dealing with our son's needs, knowing there will always be a team of people at the hospital ready with their support.

GRO-A

I am the mother of a three-year-old boy who is a haemophiliac. He was diagnosed as having haemophilia when he was one. At the time he complained about his left leg hurting and he would not walk, so I took him to the doctor, who said he had fractured his femur. I had to take him to the hospital for an X-ray, but nothing was broken. While I was there, the nurses kept coming in one by one and looking at GRO-A in a very strange way as he had rather a lot of bruises up his legs and forearms. Then the Sister came in on her own, shut the door and accused me of abusing GRO-A

I could see he had a lot of bruises, but I thought it was normal for a child of his age, just beginning to walk and falling over all the time. I was very upset about being accused of battering my baby and especially by someone I worked with when I was nursing. I saw the paediatrician who was very nice but worried about the bruises on GRO-A and asked if there were any bleeding disorder in my family. There is – for my sister and mother have Factor VIII deficiency. So, after blood tests, GRO-A was diagnosed as having haemophilia.

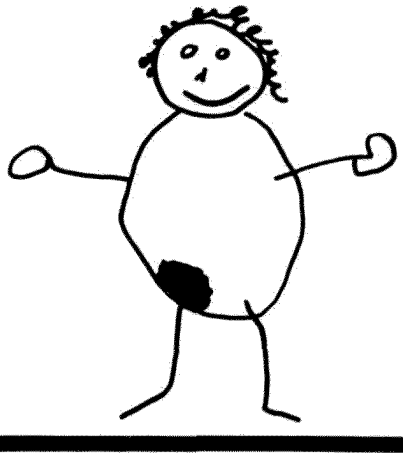
NORMAL

GRO-A is now a very active normal four-year-old who goes to a normal playschool. We have a few problems with bleeding now and again, but not too often. He is aware of his bleeding disorder and he tries to be careful, but I find it very difficult telling him that he cannot play rough games, as he wants to join in. I try not to stop him doing these things, I just warn him to be careful, or we will end up in hospital.

I have to take him to the local hospital when he needs treatment which usually means a very long

Through the eyes of a child

This is my little brother, GRO-A by GRO-A (5)



From previous page

wait of two to four hours and a few doctors all looking at GRO-A and not really sure what to do. Haemophilia is quite a specialised subject and some paediatricians are not very confident in dealing with these children. They always have to ring the Haemotology Consultant and tell him that GRO-A is here at the hospital and should they treat him? Eventually GRO-A gets his factor VIII and we go home.

GRO-A is now beginning to accept he has to have treatment when he has a bleed and he also tells me that it is hurting before we can see anything. He also sits still now when he is being treated, whereas before he used to struggle and cry and get very distressed.

We go to see a specialist every four months, as he likes to know about any problems we have had over the past few months, to air our views about any other matters we may be worried about. I find this visit very valuable as I am able to say anything I want to about GRO-A and there is support from the Dentist, Social Worker, Nursing Sister and Paediatric Consultant who give their full support in caring for GRO-A They also give him an examination all over, so they put your mind at rest and reassure you that your child is just as normal and healthy as the next child.

SKYDIVE MARK HAS HAPPY LANDINGS

PARACHUTE JUMP BY WELSH TRIO RAISES £425

It was a giant leap, never mind step, for woman as well as mankind when 22-year-old GRO-A from Monmouth in Gwent joined with his pal GRO-A nd girlfriend GRO-A in a 2,000 ft parachute drop over Herefordshire.

The sponsored event, well subscribed to by GRO-A and GRO-A 's respective families and friends, raised £425 for the Society's South Wales group's funds.

GRO-A was very taken with 5ft-nothing GRO-A bravado, the jump, he said, "having taken a lot of guts". GRO-A a cashier with a local building society.

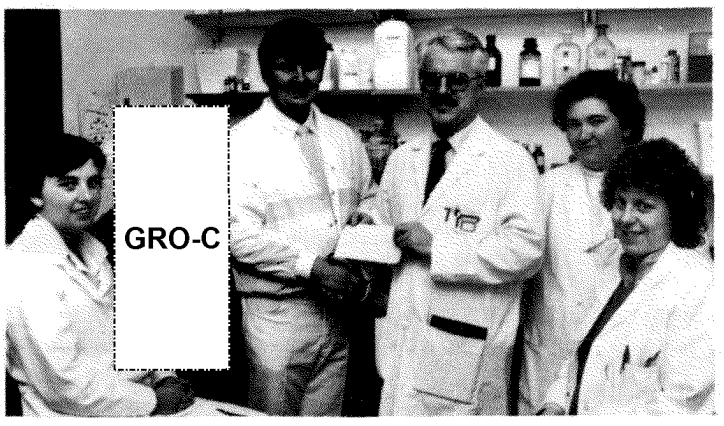
An active fund-raising member of the South Wales group, GRO-A was equally proud of his buddy GRO-A , a carpenter, who's the same age as himself. GRO-A is currently studying hard for his A levels.

Other monies raised by the local group include £64 from a courtesy transport service, organised by the Chairman, Mr Stan Slater and his son Paul. Stan has also received a cheque for £500 from the University College of Wales, the Society being one of 18 charities to have benefited from fundraising activities of the students last year.

GRO-A

OIL COMPANY'S SCOTTISH DONATION

Grampian Region Haemophilia Treatment Centre at Aberdeen Royal Infirmary has been given £150 by platform workers with the Conoco oil company.



In the photograph Ralph Reader, Chairman of the Charity Committee of Hutton TLP, a Conoco subsidiary, presents Dr Bruce Bennett with a cheque representing the sum of the fund-raising efforts of the Lasalle Crew, working in the Moray Firth. The money came from a disco evening.

Also pictured is GRO-A aged 13, the son of the Society's Aberdeen representative, Mrs Carol Robertson. The ladies shown are all laboratory assistants at the Centre.

★ *Wishes can come true..... GRO-A (left) and his friends GRO-A and GRO-A ongratulate each other after the jump. See also front page.*

FACTOR VIII INHIBITORS IN HAEMOPHILIA

by Dr. J. Beard M.B., M.R.C.P. and Dr G. Savidge M.D.

IN COAGULATION the term "inhibitor" is used in the context of both physiological and pathological states. There are several naturally occuring inhibitors in the blood which are needed for the regulation of the normal haemostatic mechanism in order to prevent uncontrolled coagulation within the circulation. In the context of haemophilia, however, an inhibitor is an acquired antibody that neutralizes the function of factor VIII.

The inhibitors arise because the patient's immune system recognises the human factor VIII infused for treatment of bleeding episodes as foreign. This results in the production of specific antibodies to factor VIII clotting activity. These antibodies develop in about 10-15 percent of multiply transfused haemophiliacs. Little is known about the basic mechanisms of factor VIII antibody production in haemophiliacs and it is apparent that the appearance of inhibitors can occur either after a few exposures to factor VIII (with approximately 40 cumulative exposure days of treatment) or much later (after 200 cumulative exposure days). Thus it is not unusual to detect the appearance of an inhibitor in the haemophilic child or adolescent.

Inhibitors are not an exclusive feature of severe haemophiliacs and, in 10-15 percent of cases, may arise in mild or moderate cases. However, in such patients they are usually transient and of low level.

In clinical practice the appearance of an inhibitor in a haemophilic patient is a serious complication. Although there is no increase in the number of spontaneous haemorrhages, when bleeding does occur it is less responsive to treatment with factor VIII concentrates as the factor VIII is rapidly neutralized by the antibody. As a consequence, even mild haemorrhagic symptoms may develop into life-threatening events. Common features of the development of an inhibitor are the appearance of seemingly therapy-resistent target joints and failure of home care programmes even with increased doses of factor VIII.

ASSESSMENT

The laboratory assessment of such patients depends on the use of screening tests for detecting the presence of an inhibitor and specific assays for determining the amount of inhibitor present.

The activated partial thromboplastin time (APTT, KCCT) is used as a screening test. A prolonged APTT may be due to either the absence of a clotting factor (such as the lack of factor VIII in a haemophiliac), or to the presence of an inhibitor. To distinguish between these two, the patient's plasma is mixed with normal plasma and the APTT repeated. Clotting factor deficiencies are corrected by the addition of normal plasma and the APTT will become normal. If the APTT is not corrected by the addition of normal plasma this implies the presence of an inhibitor.

The most commonly used quantitative inhibitor test is the Bethesda assay. It is based upon measuring the decrease in factor VIII activity when the inhibitor is added to normal plasma under specific conditions. One Bethesda unit (B.U.) is the amount of inhibitor which will neutralize 50 per cent of the factor VIII activity in 1 ml. of normal plasma incubated at 37°C for two hours.

In clinical practice, basal antibody titres (i.e. antibody levels in patients free of treatment for six months before the test) vary considerably in individual patients, and it is useful for purposes of classification and treatment to divide patients into low titre (<5 BU/ml) and high titre (>5 BU/ml) categories. It is also important that inhibitor titres and cross-reactivity of the antibody to porcine factor VIII material be established in each case since this animal product may be an effective therapeutic alternative to human factor VIII in the treatment of patients with inhibitors.

Although antibody titres are essential in the assessment of the inhibitor patient, it is also critical to ascertain the nature of the antibody response in individual cases to challenge with human factor VIII material. Provocation of the patient's immune system in this manner promotes an anamnestic response, i.e. a rise in antibody titre which can be detected approximately 5-8 days after treatment and reaches maximum levels after 2-3 weeks. Patients with antibody levels increasing by less than a multiple of three are termed low responders, whereas post-challenge levels in excess of ten times the basal inhibitor titre are categorised as high responders.

For practical purposes, intermediate responders (three to nine times basal level) are usually included in the high response group. On the basis of basal antibody titre and the nature of the anamnestic response to factor VIII challenge, a classification of inhibitor patients may be proposed and used to guide rational therapeutic policy for the management of bleeding symptoms.

PATIENT TREATMENT

Two major therapeutic aims are evident in the management of antibody patients. The primary, more short-term objective is to treat haemorrhagic episodes successfully by achieving satisfactory haemostasis. In the longer term, the goal is total eradication of the inhibitor so that the haemophiliac can become responsive to conventional factor VIII replacement again. Considerable controversy currently prevails as to the most effective treatment policy of these patients, due to the poor understanding of the mechanisms of antibody production.

There are four main methods of treating bleeding episodes in inhibitor patients: giving massive doses of factor VIII to neutralize the antibody; bypassing the inhibitor with factor IX concentrates or activated prothrombin complex concentrates (APPCs); removing the inhibitor; and using products less susceptible to inactivation by antibody i.e. porcine factor VIII. The method used depends on the severity of the haemorrhage, the inhibitor titre and the pattern of anamnestic response.

The inhibitor classification and its therapeutic application as adopted by the Haemophilia Reference Centre at St. Thomas' Hospital in London is shown here:

It is the general consensus that the treatment of choice in low titre, low responders is human factor VIII. High doses in the order of 10,000 iu. followed by 1000 iu/hr by continuous infusion are essential to overwhelm the antibody and achieve therapeutic levels of factor VIII. The decision to use either human or porcine material is determined by the patient's antibody response in each case and on the assessment of their relative merits in terms of cost-effectiveness. In some Centres plasma exchange is used as adjunctive therapy to try to reduce the level of antibody. However, this has limited application due to the transitory inhibitor depletion achieved and rebound of the antibody level after the procedure.

ARGUMENT

In high titre, low responding cases, where cross-reactivity to porcine material is low, there is a

Basal Antibody Titre	*Anamnestic Response*	*Recommended Therapy*
Low (<5 BU/ml)	*Low responder*	**Human factor VIII in increased dosage or Porcine factor VIII (if more cost effective).**
High (>5 BU/ml)	*Low responder*	**Human factor VIII (if anti-human titre <20 BU/ml) or Porcine factor VIII (if anti-porcine titre <15 BU/ml).**
Low (<5 BU/ml)	*High responder*	**Factor IX or APCCs for non-lifethreatening symptoms. Human or Porcine F. VIII for life-threatening symptoms.**
High (> BU/ml)	*High responder*	**APCCs**

very persuasive argument for the use of porcine factor VIII, due to its higher relative potency and reduced immunogenicity compared to human factor VIII. High dosages of human factor VIII are usually effective in cases where anti-human antibody titres are less than 20 BU/ml, and porcine material when the anti-porcine **antibody titres are less than 15 BU/ml. Here again plasma** exchange may be of limited value.

In high responding cases with low titre basal antibody levels, our policy is more concordant with that of Centres in the U.S.A. than with the current U.K. view. In the treatment of minor bleeding episodes in this group, the intent is to achieve haemostasis without provoking a strong antibody response. In this way titres remain as low as possible so that, if treatment is needed for a life threatening bleed, factor VIII concentrates (human or porcine) can still be used effectively at least for 5-8 days until the rise in antibody titre precludes further successful use of this material. Thus we use factor IX concentrates or APCCs for minor bleeds in these patients.

APCCs contain variable amounts of activated and precursor clotting factors. The two major preparations available are Autoplex (Hyland) and FEIBA (Immuno). The active principle responsible for their factor VIII bypassing activity is unkown. No useful laboratory tests are available to monitor their effect on blood clotting in the patient. Because of this the dose and timing of administration of these products must be determined empirically. In high titre, high responding patients, our policy relies upon **the efficacy of high dose APCCs (75-100 units/kg 6-12 hourly).**

In many Centres in the U.K., factor IX and APCCs are not advocated because of their poorly understood mode of action, lack of laboratory tests to monitor their effectiveness and their not inconsiderable high cost. These Centres use porcine or human factor VIII in all inhibitor patients, irrespective of antibody titre and response, based on clinical data that some high titre, high response patients still show improvement. Again, plasma exchange may be used as adjunctive therapy to try to reduce the antibody titre and render the factor VIII more effective.

STEROIDS

Although the use of steroids and other immuno-suppressive agents have a role to play in the management of acquired factor VIII inhibitors in non-haemophilic patients, their use in haemophilic patients with inhibitors has been disappointing.

Over the last decade, considerable interest has arisen concerning possible therapeutic approaches in the eradication of factor VIII inhibitors. The Haemophilia Centre in Bonn has **reported a series of patients where the antibody disappeared** after long periods of daily, high-dose human factor VIII therapy. In the original treatment protocol 200 units/kg/day human factor VIII were administered and APCCs used to control any bleeding episodes occurring during the programme.

For obvious financial reasons, this high-dose protocol has not been adopted in the U.K. and similar regimes applied in other countries have not been reported to have been as successful as the Bonn experience. On the other hand, low dose immune tolerance programmes have been pursued in several Centres in this country using various doses of human factor VIII on a regular basis. Although initial reports indicate that reduction of inhibitor levels and, in some cases, antibody eradication have occurred, these regimes have not been adopted on a wider scale. This may be due to the various approaches that have been used in terms of dosage of factor VIII, intervals between infusion and the general view that factor VIII inhibitor patients are, in themselves, a heterogeneous group in which no one patient may respond identically to another.

When due consideration is made to the currently known clinical **aspects of factor VIII inhibitors, it is clear that the** haemophilic patient with this complication requires the expertise and resources of a large Haemophilia Centre to ensure successful management. It is, unfortunately, all too often that attempts to treat these patients in smaller units lead to disastrous results which, in many instances, could have been avoided by prompt referral.

To some extent, this may explain the observation that only half of the registered inhibitor patients in the U.K. regularly receive treatment, and it is regrettable, but true, that many patients are of the opinion that bedrest at home is potentially less dangerous than the therapeutic attempts of haematologists in small centres with little specific expertise in inhibitor patient management.

Dr Beard and Dr Savidge are based at the Haemophilia Centre, St. Thomas' Hospital, London.

CONTACT LIST FOR LOCAL GROUPS

In order to establish contact with your local Group you should WRITE in the first instance to the Group which appears to be nearest to you – your local Haemophilia Centre will also have knowledge of your local Group and the contact person there.

BIRMINGHAM:
Mr T Fitton
42 Florence Road
Sutton Coldfield
West Midlands B73 5NG

CAMBRIDGE & DISTRICT:
Mrs C Brooks
Old Close, Hockerhill School
Dunmow Road, Bishop's Stortford,
Herts CM23 5HX

COLCHESTER:
Mrs J. Bleeze
68 Cloes Lane
Clacton-on-Sea
Essex

DERBY:
Mrs K Hill
Haematology Department
Royal Infirmary
London Road
Derby DE1 2QY

EAST KENT:
Mrs P M Clark
12 The Larches
Higham, Rochester
Kent ME3 7NQ

HAMPSHIRE:
Mrs M Clark
48 Hewett Road
North End
Portsmouth
Hants PO2 0QP

JERSEY:
Mr P J Picot
'Romali'
Chemins des Moulins
St Quens
Jersey

LEICESTER & RUTLAND:
Mrs D B Gilmour
10 Aintree Crescent
Oadby
Leicester LE2 5GD

LEWISHAM (South London):
Mr GRO-A
147 Wellmeadow Road
Catford
London SE6 1HP

LINCOLN & DISTRICT:
Mrs S L Cook
'Fairfields'
Lincoln Road, Dunstan
Lincolnshire LN4 2EX

MERSEYSIDE & DISTRICT:
Mr J Lander
10 Gypsy Grove
Liverpool L18 3LH

NORFOLK & NORWICH:
Mr G Hazlewood
Wellgreen View
4 School Road, Frettenham
Norfolk NR12 7LL

NORTHAMPTON & DISTRICT:
Mr R T Emery
The Cranes
74 Sutton Street, Flore
Northampton NN7 4LE

NORTH EASTERN
Mrs M Dickason
54 Shaftesbury Avenue
The Roundhay, Leeds LS8 1DT

NORTHERN:
Mrs P Sanderson
5 Newstead Court
Glebe, Washington
Tyne & Wear NE38 7PE

NORTHERN IRELAND:
Mr J McIlwrath
62 Bav Road, Larne
County Antrim

NORTH WALES:
Mrs C Holliday
54 Bastion Gardens
Prestatyn
Clwyd LL19 7LU

NORTH WEST:
Mrs N Guy
78 Park Road, Bolton
Lancs BL1 4RG

OXFORD & DISTRICT:
Mrs R Cleworth
64 Church Road
Upper Wanborough, Swindon
Wilts SN4 0B2

SCOTTISH:
Mr K Holmes
41 Dick Place
Edinburgh

SHEFFIELD:
Mrs Judge
52 Toll Bar Road
Sheffield S12 2QZ

SOUTHERN:
Mrs E Burrows
12 Tennyson Road
Wimborne
Dorset BH21 1NT

SOUTH ESSEX:
Mrs M Baker
'Rekabs'
47 Seaforth Avenue
Southend-on-Sea
Essex SS2 4ER

SOUTH WALES:
Mrs D Frowen
16 York Place, Risca
Newport NP1 6FR

TAYSIDE:
Mr B G Bissett
18 Whinfield Road
Montrose
Angus DD10 8SL

In addition to LOCAL GROUPS, the Society also has a new structure called SOCIETY REPRESENTATIVES. At the time of compiling this information there is only one such appointment.

ABERDEEN
Mrs C Robertson
37 Cummings Park Crescent
Aberdeen
AB2 7AS

If you require help or advice with any matter, please feel free to contact the Society's office. This applies particularly to matters relating to MOBILITY or ATTENDANCE ALLOWANCES. The General Secretary will be happy to discuss any problem which you are experiencing and where necessary take action on your behalf.

THE HAEMOPHILIA SOCIETY, 123 Westminster Bridge Road, London SE1 7HR 01-928 2020.

SPORTS enthusiast GRO-A GRO-A who is 18, is keen to make penfriends anywhere in the UK.

His haemophilia aside, GRO-A s favourite sports are cricket, hockey and badminton and other than these he spends his spare time "reading knowledgeable and interesting books, watching television and listening to music . . . all except classical", his favourite topics of conversation being sports, religion and politics.

The qualities GRO-A says he is seeking in his penfriends are "a sense of humour, kindness, and honesty". If you are interested in writing to this non-smoker, who is also teetotal, then the person to contact is, once again, David Watters.

THE NORFOLK AND NORWICH HAEMOPHILIA CENTRE

"A fine city" said George Borrow (1803-1881) in *Lavengro* and those familiar with the area would wholeheartedly agree with this. Approaching the city along the infamous A11, one can see this legend on the signs at the city boundaries.

Norwich has a population of some 125,000 and was developed to a large extent from wealth brought into the area during the 16th and 17th centuries by the then thriving worsted industry. Nowadays, we have Norwich Union, Colman Foods (famous for their mustard) and the Canaries – Norwich City Football Club. Norwich is in direct communication with the sea at Great Yarmouth via the rivers Wensum and Yare.

Contrary to popular opinion, the city is not entirely flat, as might be confirmed by anyone attempting to climb Kett's Hill on a bicycle against one of our notorious easterly winds! We have a beautiful cathedral dating from the 12th century and an equally famous castle, built on a mound in the city centre and dating from Norman times. The Broads consist of a complex of wetlands to the east of the city. These were originally medieval peat-cuttings that later became flooded. *"All roads lead to Norwich"* and the nearest centre of any size is Cambridge, some 62 miles to the south-west. The rest of the county is sparsely populated and is mainly agricultural in nature; in fact the distance between our two furthest flung patients (if that is an appropriate term for haemophiliacs) is 65 miles!

The Norfolk and Norwich Hospital itself is situated within the city boundaries and comprises 780 beds, most of them acute. Although there are other hospitals in the District, the "N & N" is the only site which is equipped for the treatment of haemophiliacs.

We look after a total of 70 patients with congenital bleeding disorders. Fifty-three of these are haemophiliacs, of whom 12 are severely affected and are on regular home-treatment. Like many other haemophilia centres we have a 24-hour cover by two consultant haematologists, the Co-Director of the **Norfolk and Norwich Haemophilia Centre, Dr. A.J. Black** and myself. We also have a Sister, Celia Shilling, who not only advises on home-treatment techniques but seems to be on continual one-to-one call. Celia is a trained AIDS consellor and is currently carrying out a project on the education of haemophiliacs undergoing home-treatment.

Perhaps slightly unconventionally, we do *not* have a large team of "experts" to deal with our haemophiliacs, preferring to call in second opinions as required. Severely affected patients are normally seen every six months, in the clinical out-patient department, when they have a thorough check-over, routine blood tests, etc. We will cope with any problems that might have arisen and receive their home-treatment records. This is the opportunity to discuss any 'AIDS-related' problems, although counselling of a more delicate nature tends to take place outside the rather formal atmosphere of the out-patient department. The summer brings thousands of holiday visitors into the area, many of them having booked cruises on the Broads, although thankfully most of the haemophiliacs amongst them are well-trained, bringing their own factor VIII or at least telling us what dose they are used to receiving!

Since the arrival of **Dr. Terry Mitchell** at Great Yarmouth in 1982, a few of our patients have been "lost" from our direct care, as they are admirably looked after by him at the **James Paget Hospital, Gorleston.** The Norfolk and Norwich Haemophilia Centre was founded in 1976, a year after I arrived; previous to this, patients would be treated on a regular basis in the N & N Casualty Department by a variety of doctors with, usually, only limited experience. We have the late **Dr. Katharine Dormandy** herself to thank for the establishment of a separate Centre at Norwich, the nearest Haemophilia Centre being at **Addenbroke's Hospital, Cambridge,** 62 miles away.

There is a thriving Haemophilia Group, currently under the expert Chairmanship of **Mr. Michael Rainsford.** The Group's Committee meets every three months, alternately in either Dr. Black's or my home, and there is a busy and active social programme, including money-raising activities. Recent projects included a Rock 'n' Roll event in March 1987, and the hiring of a market stall for a day in Norwich City Centre.

The coagulation expertise in Norwich has recently been reinforced by the arrival of a Senior MLSO in the Coagulation Section who has been trained in the **Katharine Dormandy Haemophilia Centre, Royal Free Hospital.** A new hospital is planned for Norwich, the first phase of which is due for completion in 1994, and by that time it is very likely we will have a third consultant haematologist, plus some junior staff.

Dr. J. Leslie

From left to right: Julie Starkings (Secretary), Stephen Edler (MLSO), Sister Celia Shilling, Dr. A.J. Black and Dr. John Leslie.

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The Haemophilia Society advises all haemophiliacs to wear an SOS Talisman. We also advise parents that their children should wear SOS Talismans, as they are not always with Mum and Dad.

Note: SOS Talisman advise us that if you are regularly taking pain-killing drugs, you should use a stainless steel Talisman, as some of these drugs reach the surface of the skin. This can cause damage to the chrome plated version. Details of prices for stainless steel types and bracelet types are available from Keith Colthorpe at the Society's Office.

Standard Chrome Pendant. Normal price £9.50. ***Special Offer £7.60*** **(plus £1 postage).**

St Christopher Chrome Pendant. Normal Price £12.30. ***Special Officer £9.84*** **(plus £1 postage).**

Please make cheques or postal orders payable to: *The Haemophilia Society.* This Special Offer ends 30th September 1987.

NEW BLOOD UNIT
From front page

fractionation. More than 20 different therapeutic products can be made from plasma and, after Factor VIII, and Factor IX for people with haemophilia, others in greatest demand are albumin for burns victims, and immunoglobins, used to increase patients' resistance to a range of infections.

BPL Diagnostics manufacture diagnostic reagents which are used by Regional Transfusion Centres and hospital blood banks to identify blood groups.

Of the new manufacturing unit, the Society's General Secretary, David Watters, comments: "It was a very great honour to attend the opening ceremony on behalf of the Society and people with haemophilia. We all look forward to the day when the output of the Elstree plant is increased. That day is now very much closer!"