The Bulletin

Edition 31, No. 3 1981

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

Member of the World Federation of Hemophilia
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THE HAEMOPHILIA SOCIETY
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YOU HAVE THE RIGHT TO CHOOSE

Members of the Executive Committee of the Haemophilia Society are well aware that the quality of care varies greatly from one Haemophilia Centre to another. The best ones provide full, comprehensive management — home treatment for all suitable patients and prompt infusion therapy for out-patients; regular review clinics which include blood tests, medical, orthopaedic and dental examinations, physiotherapy check-ups and the help of the social worker with problems of schooling, employment, accommodation and mobility allowances.

Unfortunately, there are some Centres where infusions are provided, often after a long wait, but very little else is done to help the haemophilic patient.

The Haemophilia Society is able to give generalised advice about standards of treatment and ancillary services. It can also, in many cases, help individuals, confidentially, with guidance and information.

The Executive Committee wishes to stress that **every** patient has the right to request his own doctor or consultant to refer him to another **of his own choice**, in order to obtain a second opinion.

VON WILLEBRAND'S SYNDROME

Classical von Willebrand's syndrome (vWd) takes its name from the Finnish geneticist, Erik von Willebrand, who first recognised and described the condition. It is an inborn bleeding tendency which can affect, and be passed on by, either sex. The pattern of inheritance varies in different families as does the severity of bleeding. Most often the chances of an affected person passing on vWd to his or her children are 50:50 each time. Affected people who are thinking of getting married and having a family can be reassured that while an affected child may need special treatment from time to time, he or she will be able to lead a normal life and to attend an ordinary school. Fear of passing on this condition, therefore, need not necessarily deter such a person from having children. Unaffected members of a vWd family, whose laboratory tests are all normal, can nevertheless sometimes pass on the condition, though this is rare.

The basic abnormality is a deficiency of von Willebrand factor. This is a protein in the blood which is associated both with platelet stickiness and with factor VIII clotting activity. In normal people, after a blood vessel has been cut, the platelets flowing out in the blood stream, stick to the site of injury, clump with one another and form a plug. After this, the clotting mechanism seals the plug firmly in place. In vWd, the ability of platelets to stick to the injured vessel is impaired, and small cuts tend to ooze until pressure is applied. This manifestation is reflected in the bleeding time test. The bleeding time which is normally less than 10 minutes is characteristically prolonged in vWd. The normal amount of factor VIII clotting activity in blood varies from 50–200% and, since the vW factor is also related to this activity, people with vWd have a reduced level of factor VIII. In addition to these tests, there are a number of ways of measuring platelet stickiness and the protein of von Willebrand factor itself that contribute to the diagnosis. These we group as the factor VIII related activities.

The clinical severity of the condition which is reflected by the abnormality in the bleeding time and the low factor VIII related activities, varies considerably within the same family. Those more severely affected, for example, may have a bleeding time of 20 minutes and factor VIII related activities of 5–10%. Once one member of the family has been diagnosed the investigation of relatives often reveals other affected people, some of equal severity, but others who will be rated mild. These people may have variable bleeding times, which are slightly long on one occasion, but normal on another, and factor VIII related activities of 5 to 50%. In fact almost every possible combination of abnormalities has been found in members of a single family, so that diagnosis becomes rather tricky at times.

Characteristically, people with vWd bruise easily and tend to bleed from the mucous membranes. For example, they may have nose bleeds or bleed from the gastrointestinal tract. Girls may have heavy periods. Superficial cuts may ooze till pressure is applied. Bleeding after dental extractions may be troublesome, but is seldom threatening.

Although vWd does not, in general, affect life expectancy and should not, therefore, jeopardise the obtaining of insurance policies, people with vWd should be properly diagnosed and issued with a special medical card, giving the results of their investigations and their blood group. Bleeding can usually be controlled by an intravenous injection of factor VIII preparation which also contains the von Willebrand factor. One such preparation is "cryoprecipitate". This is produced at Blood Transfusion Centres from normal donor blood. For some types of bleeding, e.g. dental extractions and nose bleeds, the so-called anti-fibrinolytic drugs such as EACA (Epsilon Amino Caproic Acid) or Cyklokapron (Tranexamic Acid) are often helpful. These are given by mouth or intravenously. They slow down the process by which clots are normally removed. They must not be taken if there is blood in the

NATIONAL AWARDS FOR ACHIEVEMENT IN EDUCATION AND IN SPORT BY HAEMOPHILIACS

In response to a number of requests the closing date for entries has been extended to 30th October. Full details and information on how to apply appear in the last Bulletin, Edition 31 No. 2

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urine (haematuria). Recently a synthetic hormone DDAVP, has been found useful in milder cases as it raises the patient's own blood level of factor VIII and von Willebrand factor – probably by causing stored factors to be released.

Operations and dental extractions should be carried out under the supervision of a haemophilia centre. Having said this, however, it is by no means always necessary for patients to have an injection of factor VIII. For certain types of bleeding the following first-line measures should be taken:–

Superficial cuts: Apply firm pressure (over a dressing which should be of the non-adherent variety if possible) for a good 15–20 minutes. Then put on a firm bandage over the dressing.

Nose bleeds: Sit up, lean forward and press the nostrils together just below the bridge of the nose. If bleeding is controlled, keep up the pressure for at least 20 minutes by the clock. If pressure does not control bleeding, sit forward and allow blood to drip into a bowl. If heavy bleeding continues unabated for 30 minutes contact the haemophilia centre and arrange to go up for appropriate treatment. A short course of Cyklokapron is often helpful. Infection predisposes to nose bleeds and antibiotic cream, such as Naseptin, may also help. Nose picking must be avoided.

Heavy periods: Many women with vWd have little or no trouble with menstruation. If periods are heavy, iron tablets may be needed to prevent anaemia. If blood loss is really troublesome, the haemophilia centre will arrange a consultation with a gynaecologist. Pills which control the periods may be prescribed, if appropriate.

Child birth: In most women with moderate or mild vWd the bleeding tendency and laboratory tests are all corrected spontaneously during the later months of pregnancy. As long as the appropriate tests have been carried out at the haemophilia centre, those who have corrected, can safely have their babies without special cover. For those who do not correct, factor VIII should be given, or at least be easily available, at the time of delivery. For this reason it is well for such patients to be booked at a hospital with which a haemophilia centre is associated.

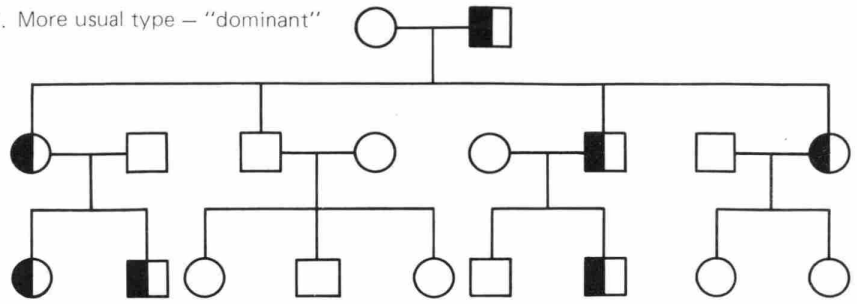
Dental extractions: Easy extractions in the less severely affected may be accomplished without undue blood loss if the socket is plugged and stitched. Cyklokapron is often given. Cryoprecipitate, other factor VIII preparations or DDAVP may be indicated.

Operations: Should be carried out at a hospital which has a recognised haemophilia centre, as it is important to raise and keep the factor VIII level at safe levels until healing is complete.

Dr. E. G. D. Tuddenham, Co-Director,
Katharine Dormandy Haemophilia Centre
& Haemostasis Unit, Royal Free Hospital

Inheritance patterns: Two typical examples:

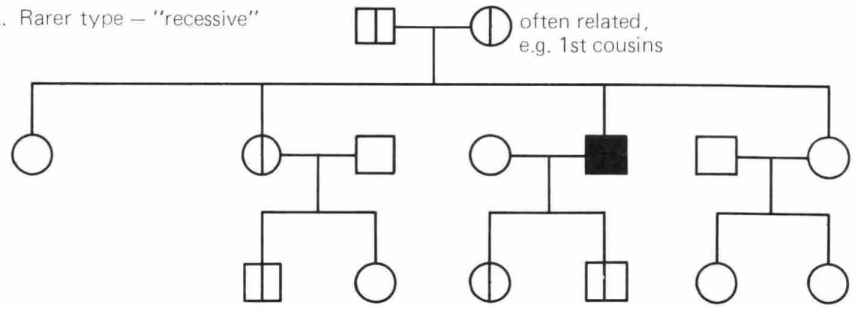
1. More usual type – "dominant"



■ Affected male □ Normal male ◐ Affected female ○ Normal female

Moderate von Willebrand's syndrome in affected members. Bleeding time 12 to 20 minutes. Factor VIII related activities 15 to 50%.

2. Rarer type – "recessive"



□□ ◐ Unaffected but laboratory tests show slight reduction of factor VIII related activities.

■ Severely affected. Bleeding time 30 minutes. Factor VIII related activities all less than 10%.

○ □ Normal and all laboratory tests normal.

BOXFILE

TRAVEL INSURANCE

We are reproducing a note below about the then new travel insurance scheme (in the name of the Haemophilia Society) which appeared in the second edition of the Bulletin in May 1979.

This special insurance for haemophiliacs themselves, family or friends, has worked well so far.

Naturally claims have occurred, but the only hint of trouble has happened on occasions when the people concerned have been somewhat "slap happy" in their arrangements.

Haemophiliacs, in almost every case, may now travel anywhere in the world. They know that all, or most, of a possible medical bill arising from an unforeseen and unfortunate incident is covered, without the usual exclusion in their case because of a pre-existing condition.

LIFE INSURANCE

The third edition of the Bulletin for 1980 contained a report on the Bonn Conference held there in October last year. Before that meeting, Mr. Rosenblatt travelled to Cologne to see leading officials of one of the world's largest life insurance reinsurance companies. Their Chief Medical Officer was persuaded to attend the whole of the Conference. The Society is pleased to announce that, as a consequence of the information he was able to obtain, the reinsurers' underwriting attitudes have been improved considerably. This will help haemophiliacs in the UK as well as elsewhere in the world, particularly in Europe.

Mr. Rosenblatt will advise anyone on life insurance matters, whether or not business is effected via his Company or elsewhere.

CAR INSURANCE

It is crucial that Haemophiliacs make their condition known to their Insurers, whether they are existing policyholders or proposing for a new policy. Failure to do this could invalidate a policy and might also prejudice a claim.

Provided you are able to show a Centre Doctor's letter confirming that you are physically capable of driving a motor car, one Insurance Company has agreed to cover haemophiliacs at standard rates. This, of course, assumes that he qualifies for standard rates in any event. Please contact Mr. Rosenblatt for any further information or advice you require.

The Haemophilia Society is paid commission by Brookdale Brealey (Insurance Brokers) as though the Society were agents. This helps swell Society funds. The arrangement applies to all insurances for Haemophiliacs, families or friends, **as long as Mr. Rosenblatt is made aware of the connection.**

N.B. Over the past two years the Society has benefitted to the tune of almost £1,000 as a result of commission earned on policies entered into as described above.

The Society is pleased to announce that a breakthrough has been made in the field of travel insurance for haemophiliacs. Messrs. Burgoyne Alford & Co. Ltd. and

Brookdale Brealey (Insurance Brokers), via Lloyds of London, in conjunction with Lloyds Brokers, have arranged a travel policy so that a person having haemophilia, or any allied disorder, can go abroad, secure in the knowledge that he will be properly covered on the medical expenses section of the policy, as well as all other sections.

There will be certain warranties and requirements but these are no more than any prudent Society member should expect. For instance, he will be required to have a letter from his Centre doctor saying that he is fit to travel. Furthermore, he should take with him sufficient concentrate to be treated, either by himself, or by someone else, should the occasion arise. Full details of these and other requirements are shown on the proposal form, which may be obtained solely from Mr. D. Rosenblatt of Brookdale Brealey (Insurance Brokers) of Valley House, Crossbrook Street, Cheshunt, Herts. Telephone No. Waltham Cross 31971.

It cannot be emphasised too strongly, that the long term success of the new scheme will largely depend on our members' not abusing it and their making absolutely certain that they make every effort to see that avoidable claims do not arise. There is no philanthropic figure in the background with an inexhaustible fund of money, who will be happily making payment of medical expenses for those haemophiliacs who have failed to make proper provision for themselves, before going abroad.

Claims paid out to members will come from a pool of money which is provided from the premiums paid by our members and their friends when taking out this new policy. The fate of this opportunity is, consequently, in our own hands.

So that the fund may be made as large as possible, members are urged to ask their families and friends, i.e. non-haemophiliacs, to effect normal travel insurance, again with Lloyds, via Mr. Rosenblatt at the firm's name and address given above. The standard Lloyds policy is competitive and has good cover. In this way the new scheme, which is of great importance to all members, will have every chance of continuing to provide insurance for haemophiliacs against unfortunate and unforeseeable illness or accidents whilst abroad.

D. Rosenblatt

Not Relevant

Not Relevant

DONOR SERVICE MAY SELL BLOOD PRODUCTS

by Andrew Veitch

By-products of blood donated through the transfusion service may be sold to drug companies.

The Health Minister, Dr. Gerard Vaughan, has confirmed in a Commons answer that talks with several firms have started. Beechams said yesterday that it was one of the companies interested.

The products, the result of processing plasma at the Government's Blood Products Laboratory, would be those which the health service does not need or does not have the facility to process, said a Department of Health spokesman.

They would not include Factor 8, used for the treatment of haemophiliacs, or Albumin, which is used to treat burns. "We cannot get enough of these and there is no question of selling them," he said.

Products for sale might include those from which treatments for tetanus, smallpox, and chicken pox are made, according to sources in the National Blood Transfusion Service.

Beechams is interested in substances which would help to develop a new treatment for deep vein thrombosis.

Dr. Vaughan said, in answer to a Commons question from Conservative MP Mr. Robert McCrindle: "I am sure that the majority of donors would wish the maximum use to be made of their donations, and so I have agreed that the department should discuss with the NHS and the companies the possible sale of such materials for the manufacture of health care products. The income from this will directly benefit the NHS."

With grateful acknowledgements to the Editor of 'The Guardian', 4th August, 1981.

Editor's Note:

Donated blood may be used either as whole blood or processed to produce a variety of therapeutic and diagnostic material. This important change should enable the maximum use to be made of all donated blood.

Not Relevant

MISCELLANY

Erratum: Edition 31, Number 2, 1981.

In the last paragraph of AGM Report we incorrectly stated that money raised in Manchester was to be used to furnish a patients' waiting room at Manchester Royal Infirmary. In fact, the money was raised by patients at the Royal Manchester Children's Hospital and will be used to help furnish a new Haemophilia Treatment room in that hospital.

MOTABILITY

You may be interested to know that Motability has been offered a special stand at this year's Motorfair at Earl's Court, which will take place between the 21st and 31st October. The organisers of Motorfair have made special facilities available, including a Motability entrance on the ground floor next to the main Warwick Road entrance at Earl's Court.

A leaflet on the Motability: Car-Leasing and Hire-Purchase schemes can be obtained from:

Motability
The Adelphi
John Adam Street
London WC2 6AZ

H.R.H. The Duchess of Kent

On 21st July 1981 the Society was represented by Dr. & Mrs. GRO-D and Mr. & Mrs. GRO-D at H.M. The Queen's Garden Party in the grounds of Buckingham Palace.

In their words: "It was a beautiful summer's day and as well as the enjoyable experience of walking around and having tea in the Palace gardens we were fortunate that the opportunity arose to have a long chat with the Society's Patron, H.R.H. The Duchess of Kent. The memory of the occasion will be cherished by us all for many years to come."

INDEPENDENCE WEEKEND

by D. I. K. Evans,
Consultant Haematologist,
Royal Manchester Children's Hospital

Five boys from the Royal Manchester Children's Hospital Haemophilia Centre went on two "independence weekends" in May and June together with three girls and four boys with rheumatoid arthritis. The idea behind the weekends was two-fold. We felt that many children with chronic diseases tend to be over-protected, both at home and at school, so we aimed to give them opportunities to do the things that they do not normally do. At the same time we hoped to see how the boys related to boys and girls with a different disease. We gave talks on home safety, personal care, and showed them Cutter Labs.' tape-slides about haemophilia. For the second weekend we had fewer talks as they were unpopular, and had a kite-making competition instead. Each group had enormous fun making and flying their kites, and the boys and girls showed unexpected agility in running up and down hill to get their kites up. The children were divided into three groups, with different diseases and sexes in each. Each group had to plan, buy, prepare, cook, eat and wash up one main meal for their group and helpers. Meals ranged from chicken curry with rice followed by banana custard to hamburgers and sweet corn. They did washing, ironing, making beds and tidying (not much of the latter!): but it was not all work. The boys played pool, table tennis and darts. The electric organ was very popular. We had a disco both Saturday nights and went swimming on Saturday afternoons as an alternative to watching the Cup Final on television for the first weekend, and badminton for the second.

The idea for the weekends came from Mrs. Valerie Mellor, principal psychologist at Booth Hall Children's Hospital. We needed a lot of helpers: some stayed the whole weekend, and others came for part of the time. They included physiotherapists Jan Davies, Eileen Warham and Emma Forsyth-Brown, Sister Alex Shaw, medical students Sam Rawlinson and Alec Mitchell, Dr. Geoff Foster and his wife Libby, Drs. John Keen, Lennox Holt and myself. We used Fourways, a residential home for young handicapped run by Wigan Social Services Department. The home was empty as most of the residents had gone home for the weekend. The expenses were met by the boys' local Education Committees and by the North West Branch of the Haemophilia Society. We are very grateful to everyone whose contributions helped.

The boys thoroughly enjoyed themselves. On the first Friday night they were all very excited and restless, but by Saturday they were exhausted and soon dropped off to sleep. Two gave themselves injections in front of an admiring crowd and enjoyed showing how well they managed. We were all impressed by the speed with which they made friends and settled in together. Some seemed to behave in quite a different way from what their parents had led us to expect. They all compared the way their parents dealt with the disease; and they often blamed their parents for limiting activities without realising why. All the children thought it was a holiday and want to go again; but we now realise how much some other children who did not come would benefit.

THE ROLE OF THE SPECIAL SCHOOL FOR HAEMOPHILIACS

by Sister P. Turk,
Lord Mayor Treloar College,
Alton, Hampshire

The role of the special school for haemophiliacs has always been a controversial subject, even more so since the widespread practice of home therapy, which has enabled many haemophiliacs to attend local schools with reduced loss of school attendances.

Nevertheless, there are still many more haemophiliacs who have, for a variety of reasons, been deprived of a normal schooling.

Treloar College provides medical and educational facilities for handicapped children including haemophiliacs and I am here to tell you of the work done at the college and how this affects the haemophiliacs in our care.

Lord Mayor Treloar College was the brain child of William Purdie Treloar, Lord Mayor of London at the turn of the century. He was appalled at the suffering of the crippled children in the poor areas of the city and used his influence to launch a national appeal to provide medical care and educational facilities for these children.

He bought a disused hospital at Alton, a small farming town in rural Hampshire which had been built for the soldiers returning from the Boer War, and thus the first hospital school in the country was opened in 1904.

When the National Health Service was formed in 1948, the College became a registered charity and moved to its present site and the original hospital school became known as Lord Mayor Treloar Hospital, now the Regional Orthopaedic Centre for Wessex. The College then provided boarding school education for physically handicapped boys from the age of 14 years.

In 1950 a sister school was built nearby on the outskirts of the town for similarly affected girls.

Over the years the types of handicaps have changed – initially the most crippling disease was tuberculosis and then poliomyelitis. In 1970 boys suffering from haemophilia were admitted to the College and Treloar Haemophilia Centre was formed at the hospital, principally to serve the needs of the College, but also to provide facilities in the area for out-patients.

During the last 10 years the haemophilia unit has grown from a modest side ward in the paediatric unit to a full and busy department including a separate haemophilia laboratory, 3 medical officers and 4 nurses catering for the needs of 40–50 college pupils and 25 out-patients.

Two years ago, the girls and boys schools amalgamated into two co-educational establishments of junior and senior schools. At the same time the haemophilia unit moved into the College premises of the senior school sick bay.

At present there are 300 children in the 2 schools, of which the present number of haemophiliacs is 42.

The educational fees for the pupils are paid by the local education authorities and the National Health Service funds the haemophilia unit and the salaries of the medical and nursing staff. All other expenditures are met from the charity trust fund.

The College provides boarding education up to university standards; most pupils enter the College at the age

of 12, but in special circumstances the entry age can be as low as 8 years.

In addition to the traditional education, the College offers a variety of practical and specialised courses, e.g. tailoring, computer studies, business and secretarial courses and horticulture. Approximately 20 of the senior pupils attend the local 6th form college which has special facilities for the physically handicapped. Classes in the college are essentially small, and the classrooms purpose-built with desks and benches at varying levels and extra wide ramped doors for ease of access for wheelchairs and trolleys. The use of electric typewriters, tape recorders and possum machines are readily available where necessary.

In addition to the educational studies, a wide variety of sports are encouraged however handicapped a pupil may be. Swimming, canoeing, archery and gymnastics are catered for on the premises and nearby facilities for sailing and horse riding are available.

Accommodation for pupils is also purpose-built – some have single rooms, but most are in twin-bedded units. As the pupils progress through the school they move out of the main building into a hostel which has its own common rooms and kitchen. Also there is an independent living flat where pupils spend 3 weeks doing their own catering and cleaning.

As well as the teaching staff, there are teams of physiotherapists, and remedial gymnasts, house parents and care staff and an occupational therapist, plus an 18-bedded sick bay covered by qualified nurses 24 hours a day. Visiting staff include a paediatrician, orthopaedic surgeon, dentist, optician, dietician and genetic counsellor.

Socially, the children are well-known in the local community and integrate with the town activities. Within the College, recreation is varied, the regular discos being popular with pupils and staff alike. There is a thriving orchestra, folk groups and drama section – in fact, many pupils complain that they have not enough free time.

As I previously mentioned, there are 42 haemophiliacs at the College – all are severe, i.e. those with less than 2% Factor VIII. Over half of these children have other additional medical or social problems. They come from as far afield as Dublin and Bristol and all points between.

Breakdown of the Present Problems

7 children are either one-parent families or under the care of the social services.

4 have behaviour problems.

7 have inhibitors – 4 low, 3 high.

Additional medical problems are as follows:–

2 cerebral dysfunction following brain haemorrhages.

3 have a degree of paraplegia following spinal bleeds.

2 have had severe internal bleeds.

1 has Perthes disease.

1 has cystic fibrosis.

2 have epilepsy.

Some of these children fall into more than one category.

On entry to the College, all haemophiliacs are screened for inhibitors and a full blood chemistry is performed and repeated every term thereafter.

A full physical examination is undertaken including detailed measurement of all joints and ranges of movements. They are then seen by the orthopaedic consultant who visits the College each week and a programme of physiotherapy, hydrotherapy, splintage or prophylaxis is initiated.

All haemophiliacs are taught from entry into the College the importance of early treatment for bleeding episodes. Thus, day or night they report to the sick bay and are seen immediately by the medical staff. All bleeding episodes are seen and treated within one hour of the first symptoms occurring.

Each morning the review clinic is held from 8.30 a.m. – 10.30 a.m. approximately. This clinic, and the treatment of fresh bleeds are the only occasions when schooling may be missed. All other clinics and treatments are done in break-times or after 4.00 p.m. when formal lessons have finished.

Following a bleeding episode the patient is reviewed daily at either morning or evening clinic until the original range of movement has been regained.

Within the sick bay area, there is a 10-bedded admission ward for haemophiliacs unfit to return to school. One of the teaching staff is on duty throughout the day, thus lesson continuity is maintained.

Plaster of Paris or Baycast splints are made in the sick bay and there is also a central store for crutches, wheelchairs, leg and footrests.

Block leather splints are made by a local firm who visit the College weekly.

Research

Because of the large number of haemophiliacs at Treloars, it has been possible to study in detail bleeding episodes in joints and muscles.

From these studies various papers have been published regarding the dose requirements for bleeds of varying severities and sites. This work is of particular importance, since the advent of home therapy has made it difficult for centres to keep a watchful eye on dose regimes.

Also major clinical trials have been carried out on various prophylaxis regimes in order to establish the optimum minimal dose required to reduce bleeding episodes.

Prophylaxis

Prophylaxis is used extensively at Treloars for a variety of reasons:

The most important use is for the treatment of target joints (i.e. joints liable to damage from recurrent bleeds into the same site). Prophylaxis is given on alternate days and during the period of prophylaxis, which is reviewed every half term, intensive treatment of that particular joint is undertaken, usually in the form of hydro or physiotherapy and night immobilisation in a block leather splint.

Short-term prophylaxis is used when the school examination periods occur, particularly those pupils who are sitting 'O' and 'A' level examinations since these exams have to be taken on the stated day and cannot be deferred.

Also short-term prophylaxis is given when long journeys are undertaken, thus half-term and end of term holidays are particularly busy times.

Surprisingly, we also give prophylaxis prior to discos, visiting rock concerts or football matches. This may seem rather irresponsible, but in hindsight a transfusion before any of these events often uses less material and certainly reduces the incidence of bleeds following these activities.

An intensive self-therapy training scheme has been practised at the college for some time which has aroused great interest from visitors to the unit. Each term 6 pupils are selected to undergo training. These training sessions are held out of school hours thus avoiding any interruption in academic studies and last ½–1 hour, each pupil being trained individually and then later on in the programme in groups.

Prior to training, the Centre director concerned is contacted and permission to commence training is obtained along with information on the particular material the centre will be able to supply during holidays. A record of this information and the progress made throughout the training is maintained and this information is sent to the Home Centre on completion of training.

Practical sessions consist of:- -

1. Personal hygiene.
2. Setting up and preparing transfusion materials.
3. Self-administration of intravenous infusions.

Before completing the practical sessions, all the pupils must have several sites for transfusions, including the feet particularly suitable when elbow bleeds occur or when the veins of arms and hands are of poor quality.

The theoretical sessions are as follows:–

1. General understanding of their condition and allied coagulation disorders and the effect on joints and muscles when bleeding episodes occur.
2. The risks and precautions of each preparation and a general under-

standing of the various materials available.

3. How to deal with reactions and the types of bleeding episodes requiring hospital assistance, e.g. severe trauma, haematuria and abdominal bleeds.
4. Documentation of bleeding episodes.
5. All trainees are taught to calculate the dose of material required. These calculations have been assessed as a result of the clinical studies carried out at Treloars as follows:—

   10% raise for ankle bleeds
   20% raise for all other joint bleeds
   30% raise for muscle bleeds.

   The dosage is calculated as follows:—

   Kg. body weight x desired rise (if over 60 Kgs divided by 2)
   1.5

   By converting this onto a graph, it is possible for all pupils to have their own copy and calculate exactly the number of units required for all bleeding episodes.

Conclusion

I hope I have given you an insight into at least part of the Treloar College scene. Our aim is to improve the quality of life of the handicapped child, certainly where the haemophiliac is concerned and results are promising.

With the ever improving treatment of haemophilia and a good standard of education, most of the pupils are able to be placed in employment or in further educational establishments, even in the light of the recent depression in employment.

WORLDWIDE
W.F.H. News

Extracts from the Bonn 1980 CONFERENCE REPORT
by GRO-D

The main thrust of Conference was divided into 3 main areas, each area covering specific aspects of Haemophilia concern, and discussed under the title of Workshops.

WORKSHOP I:
"COMPREHENSIVE CARE PROGRAM FOR HAEMOPHILIACS"

The recommendations which emerged from the workshop session and presented to Congress for implementation were as follows:—

1. Haemophilia referral centres should be established within each country and comprise a basic care team of Haematologist, Orthopaedist and Physiotherapist, as well as Dental Support Staff.
2. Each Centre should establish a specialist nursing capability, including nursing training and information, and support Social Worker input, trained in the special problems of Haemophiliacs.
3. Enhance the referral of all Haemophiliacs to such Centres and establish close co-operation between the elements of a Network of such Centres, especially with regard to the collection and dissemination of clinical and statistical data to better utilise existing treatment and care resources.
4. Encourage and promote the availability of supervised self-treatment facilities to all suitable patients.

WORKSHOP II:
"MOBILISING SUPPORT FOR HAEMOPHILIA CARE"

This workshop session divided into two separate areas for discussion.

i. Maintenance of the Plasma Supply and Plasma discussion.

ii. Financing the cost of Haemophilia Care.

The recommendations emerging from Workshop II to Congress for task force implementation were as follows:—

1. Promote the availability of sources of fresh frozen Plasma and to encourage technological development to improve fractionation yields.
2. Promote research into the improved handling and packaging of high yield materials such as Cryoprecipitate to the point where they may be suitable for Home Therapy Application.
3. Strive to encourage national self sufficiency in plasma production.
4. Each country to establish effective and equitable treatment funding including equitable pricing of commercially produced material.
5. Ensure adequate guidance and assistance be available to countries to assist in setting up the necessary mechanisms for plasma production and fractionation, especially in the 3rd World.
6. Encourage 3rd World country Blood Transfusion Services to develop high volume cryoprecipitate production.
7. Develop and strengthen inter agency collaboration, e.g. World Health Organisation etc., in relation to 3rd World development.

WORKSHOP III:
"NATIONAL HAEMOPHILIA SOCIETIES AND THE WORLD FEDERATION"

The main criteria governing the role of societies at all levels, apart from the supporting of vital scientific, clinical and social research, and the role of pressure groups, was the need to

1. inform
2. be informed
3. be able to resolve specific problems.

In order to achieve this role, national bodies will require to encourage and develop strong regional and local self help groups integrated into the National Society structure in a meaningful and participant way.

The recommendations from Workshop III to congress task forces were as follows:—

1. Support the establishment of WFH information clearing house by encouraging submissions from all relevant sources and material.
2. Promote the establishment of Self Help groups supporting National Societies.
3. Strengthen the liaison between societies and researchers at all levels.
4. Support continuing program of education in Haemophilia throughout the World.
5. Support and encourage the involvement of young people at all national and international levels.

Some state of the art recommendations from Haematologists and Orthopaedists in the treatment of Haemophilia were given to Congress as follows:—

1. Replacement therapy should not be withheld even if no obvious clinical signs of a bleeding episode are manifest, even to a trained observer.
2. All patients should have access to self-treatment programs and where suitable, should be encouraged.
3. All surgery should be supervised by trained Haematologists.

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THE XIVth CONGRESS AND XVth GENERAL ASSEMBLY OF THE WORLD FEDERATION OF HEMOPHILIA

I attended the XIVth Congress of the WFH, held in San José, Costa Rica from 3—7 July, 1981, on behalf of the Haemophilia Society, and acted as the UK voting delegate at the XVth General Assembly. This report comprises, firstly, an account of the General Assembly, followed by notes on other aspects of the Congress.

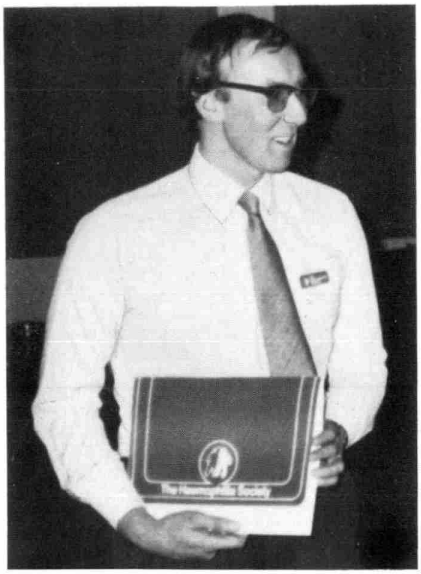
1. **XVth General Assembly (Sunday 5 July, 1981)**

   (a) *Credentials Committee*

   It was stated that all National Member Organisations present were regarded as being in good standing and qualified to vote. The constitutional position of countries which are, or have been, in arrears still seems unsatisfactorily obscure.

   (b) *New National Member Organisations*

   Paraguay and Honduras were elected members of the WFH.



Ken Milne

(c) *Minutes of Bonn XIVth General Assembly*

These were approved, despite many delegates (including UK) not having seen them.

(d) *World Hemophilia Youth*

The Annual Report of WHY was presented by GRO-D (Canada). WHY now has a mailing list of more than 180 in 38 countries, and national/regional organisations have been set up in Canada, USA, UK, Sweden and Malta. WHY intends to be involved in the following projects:—

(i) Production of a 3rd Edition of the Guide for Travelling Hemophiliacs.

(ii) Involvement in the Hemophilia Action Group, in particular in Task Force III.

(iii) Direct assistance to haemophiliacs in developing countries. It is planned to include a youth programme at the 1983 Congress in Stockholm. Such a programme was held in San José. GRO-D will be reporting on WHY in greater detail.

(e) *WFH Information Clearing House*

The Clearing House in Heidelberg was opened last October and is now in operation. More details are given under "Task Force III" below.

(f) *International Hemophilia Training Course*

The 1980 report from Professor Mannucci, the IHTC Chairman, was included in WFH Bulletin No. 18. Training fellowships were granted to 6 people during 1980. No Workshops were held in 1980, but future workshops are planned for 1982 (Jamaica) and 1983 (Philippines).

(g) *Home Care Committee*

The Home Care Committee has sent over 6000 questionnaires to samples of haemophiliacs in 30 countries. Only incomplete returns have as yet been made, however, although results from 2 countries have been analysed. A study has also been made of Inhibitor patients on home care.

(h) *Hemophilia Action Group*

Brief details were given of the work undertaken by the Task Forces set up to implement the recommendations of the Workshops held at the Bonn Conference. The Task Forces are referred to later in this report.

(i) *World Health Organisation*

The Executive Board of the WHO has undertaken its triennial review of non-governmental organisations in official relations with WHO, and is to continue to maintain official relations with WFH. It is intended to draw up a framework for activities to be jointly undertaken by WFH and WHO during the next 3-year period. In 1980 the WFH was represented at WHO meetings in Geneva, Malé, Manila and Washington.

(j) *European Advisory Board*

John Prothero, our European Liaison Officer, expressed disappointment at the lack of progress by the EAB in the last year, but hoped that an EAB meeting would be arranged in Holland in November.

(k) *Costa Rica Prototype Project*

The development of the Costa Rica project was reviewed, and the Assembly offered its congratulations to Dr. Cordero on the success achieved in the ten years since the project was started in 1971. Certainly from the evidence of the papers read at the Congress and from the visit to the Hospital Mexico, the project has been very successful and could be emulated by the more affluent third world countries. However, the success has been achieved only as part of a general campaign to establish a national health/social security programme, and this programme clearly draws on a large proportion of Costa Rica's resources. Whether such a programme could be undertaken by the poorest ("fourth world") countries must be rather doubtful.

(l) *International Year of Disabled Persons*

It is hoped that the IYDP can be used to improve haemophilia care, and particularly to encourage development of rehabilitation programmes for haemophiliacs.

(m) *Financial Matters*

The Auditor's Report for the year ended 30.6.80 was presented. A budget for 1982 was presented, indicating an estimated income of US$138,000 and estimating expenditure at $130,000. Income from National Member Organisations (assessments plus arrears) is estimated at $27,000. The Assessment Schedule for 1982 was approved. The assessment for the UK for 1982 will be $2,040. For comparison, the six other countries contributing more than $1,000 are:—

USA	$11,120
Japan	4,260
Fed. Rep. Germany	3,710
France	2,820
Italy	1,610
Canada	1,530

It was emphasised that cash flow problems at the WFH made it important for payments to be made as early as possible, and that agreement to do so was implicit in approval of the assessment schedule.

(n) *Future Congresses*

The venue and date of the XVth Congress were approved — viz. Stockholm, Sweden; June 27—July 1, 1983. One of the proposed topics outlined, and of particular interest to us at the moment, is that of international blood supply. The Congress will include a youth programme, and, in view of the high cost of living in Sweden, there will be facilities for low cost accommodation, e.g. camping. The Congress will immediately precede the IXth International Congress on Thrombosis and Haemostasis, also to be held in Stockholm. The venue of the XVIth Congress was approved as being in Brazil (either Sao Paulo or Rio de Janeiro), as offered by the Brazilians in Bonn, in either 1984 or 1985. The date is dependent on the decision of the WFH Executive as to whether the linkage of the Congress with other Scientific meetings should be changed.

2. **Attendance at XIVth Congress**

The Congress was well attended, naturally enough, by the Latin American Countries, to whom the general theme of haemophilia care in the developing countries was especially relevant. The European countries were less well represented, the absentees including Austria, Belgium, Eire, Finland, Switzerland and the Iron Curtain countries.

3. **Poster Presentation**

Each National Member Organisation attending presented maps relating to the geography of haemophilia, it being intended to publish these with the National Status Reports presented in Bonn. While the quality of the maps presented was very high, the presentation was less valuable than that in Bonn, in that no specific session was allocated to it. Consequently people tended to visit it when they had a few spare moments, and the personal communication which was such a successful feature of the Bonn Conference was not practicable. However, I was able to talk usefully to a fair number of people, and our pamphlets etc. were again much in demand.

4. **Task Force I (Care for the Haemophiliac)**

The recommendations of Bonn Workshop I were received (see WFH Bulletin — Bonn Special Issue).* A Nurses' Working group has been established, chaired by Christina Brackmann (Bonn), and a report has been produced defining the (ideal) qualifications and duties of the Haemophilia Centre Nurse. The working group has also produced similar reports, in draft form, for Social Workers and Physical Therapists in Comprehensive Care Centres. Discussion mostly centred on the relevance of the work of the Task Force to less developed countries, it being alleged, with some justice, that its work so far relates

to the highest standards of care achievable only in highly developed nations, and that its work might give the impression that haemophilia care is only for the rich. Dr. Brackmann's view, however, was that it is necessary to establish optimum standards first, and that only then can it be considered to what extent these standards can be diluted to suit circumstances in particular countries.

* A Summary of Bonn Workshop I recommendations appears on page 6 of this Bulletin.

5. **Task Force II (Support for the Haemophiliac)**

The recommendations produced in Bonn regarding supply of treatment materials were discussed. The Task Force has been mainly involved in fact-finding since the Bonn Conference, and has co-opted a number of people to look into specific aspects of the problems. The people responsible for each aspect are:—

Tony Britten (USA)	— Plasma Supply
Franz Etzel (F.R.G.)	— Fractionation
Dr. Smit-Sibinga (Holland)	— Cryoprecipitate production
Feredoun Ala (UK)	— Standardisation International movement of material
Charles Carman (USA)	— Pricing Payment

An implementation plan for Task Force II was presented, as indicated below.

Strategic Goal (1980–1990)

To stimulate the production of fresh frozen plasma for fractionation and cryoprecipitation, throughout the world, to ensure adequate availability of factor VIII for haemophilia care.

Short-term Objectives (1980–83)

Stimulate increased production of fresh frozen (source) plasma, throughout the world, to ensure increased source material for factor VIII production.

Stimulate the development of high-yield factor VIII production, by expert fractionators, to encourage the increased availability of factor VIII concentrates.

Stimulate the development of cryoprecipitate production capability, in every country, to ensure accessibility to treatment for all haemophiliacs.

Strategic Goal (1980–1990)

To stimulate the establishment of effective methods of paying for haemophilia treatment, throughout the world, to eliminate artificial financial barriers to haemophilia care.

Short-term Objectives (1980–83)

Work through all available channels, in all countries, to establish mechanisms for payment for haemophilia treatment.

Encourage national haemophilia societies to take appropriate action against unfair pricing, in each country, to ensure fair and equitable market prices.

It has not yet proved possible to commence the multi-centre study of dosage levels which was agreed in Bonn. The work of Task Force II is of very great relevance to our own present concern

Not Relevant

with Blood Products, and this session was, in my view, the most directly valuable to our Society. I propose to keep in touch with members of Task Force II concerning Factor VIII Supply and Demand.

6. **Task Force III (Information and Organisation)**

Once again the Bonn recommendations were reviewed and discussed. The major item for discussion was the Information Clearing House in Heidelberg. The Clearing House has already produced a list of literature references received since October, 1980. The Director, Professor

Not Relevant

Schimpf, is, however, disappointed at the limited response to his request for literature, only ten countries (including the UK) having sent material. The question was raised as to whether reprints of articles could be supplied from Heidelberg, but Professor Schimpf felt this would be too expensive.

7. **Medical/Scientific Sessions**

I was unable to attend many of these sessions, as many were simultaneous and I felt it desirable to give priority to attending the Task Force Sessions. Among the subjects of particular interest to us were:—

(a) The use of Prothrombin Complex Concentrates in haemophiliacs with inhibitors. "Autoplex" (Hyland) seems promising, and among those reporting its successful use was Tony Aronstam (Treloar College).

(b) The use of Porcine Factor VIII Concentrate ("Hyate C" – Speywood) in treating inhibitor patients (reported on by Peter Kernoff of the Royal Free Hospital). The polyelectrolyte fractionation technique used in producing "Hyate C" may also be of considerable value in producing human Factor VIII, offering the possibility of greatly increased yields.

(c) Prenatal diagnosis. Although we are already familiar with this subject, it is worth noting the great interest expressed in it, and Dr. Mibashan's failure to find anyone prepared to express doubts about its ethical validity (at a Congress in a Catholic country well attended by Latin-Americans!). There was an interesting paper about the socio-ethical consequences of prenatal diagnosis, with the suggestions that guilt feelings are more common among those who undergo abortion for genetic reasons than among those those whose abortions are for other reasons. It was suggested that increased support for families in this position is needed.

Many of the papers presented related to development of haemophilia care programmes in developing countries, and consequently their content was not of direct relevance to the situation in the UK.

8. **Future Format of Congresses**

The San José Congress, like the Bonn Conference, was intended to offer parity between scientific and social aspects of haemophilia. An eloquent and able speech was made by the Spanish delegate at the General Assembly requesting that yet more emphasis be placed on social, or socio-medical aspects. This view seemed to be supported by the Latin-American nations. No firm conclusion was reached on this suggestion, but if implemented the support received from medical participants might diminish, as it might if the link with the ICTH is broken.

GRO-A

Ken Milne

Opinions expressed in the Bulletin do not necessarily reflect those of the Haemophilia Society.