The Bulletin

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MAJOR CHANGES AT THE SOCIETY'S NATIONAL OFFICE

The Society has undertaken a major restructuring exercise to shape it for the challenges of the year 2000 and beyond.

This has taken place as a result of the Society's work in implementing its Strategic Plan. The planning began over two years ago when priority areas were identified. Having identified the priorities, the Executive Committee considered the structure of its own internal working procedures and reviewed its national office staff and premises.

At the same time an extensive needs assessment exercise involved the membership in identifying the changing requirements of people with haemophilia.

The Executive Committee has now implemented the restructuring of the national office organisation in line with the needs of the Strategic Plan.

The national office now operates with two departments which reflect the new Executive Committee structure. The Services Department, under Graham Barker handles the core work of the Society. This work includes such items as services to individual members and groups, campaigning and government and public relations. The Resources Department under Susan Archer deals with the day to day running of the Society office, such as office management, finance and membership administration. Graham and Susan should now be your main points of contact at the national office.

"The two department format, with each dealing with separate aspects of the Society's work is the outcome of the review of our approach," said Haemophilia Society Chairman, the Revd Alan Tanner.

"The new structure will enable us to continue to be at the forefront in matters that affect the lives of people with haemophilia and yet have a greater flexibility to involve a wider cross section of the membership in the operation of the Society." Having the two departments, providing mutually complementary services, means that the Society has greater depth of resource available to provide services for people with haemophilia."

The new structure has meant that the Society has had to lose some existing posts, while creating two new positions at the national office. One of the posts that has been lost is that of General Secretary of the Society.

"David Watters, gave the Society sterling service over the years and successfully led the organisation through a critical period when the campaign for compensation was high on our agenda," said the Revd Tanner. "We wish him every success in the future."

Two new posts have already been advertised and appointments are expected soon. The posts are Member Services Officer and Head of Fundraising. Both are seen as crucial to the success of the new structure. The Member Services Officer will support the work of the Services Department, allowing a greater depth of resource to be available. The Head of Fundraising will also have a vital role to play. The Society will need to at least double its annual revenues by the year 1995 to finance its planned expansion in services.

KEN MILNE

It is with the deepest regret that we report the death of Society Vice Chairman Ken Milne.

Ken was a long serving and staunch supporter of the Society and he will be greatly missed.

A full tribute to Ken will appear in the next edition of The Bulletin.

TRINIDAD & TOBAGO

GRO-A

Officials of the Society for Inherited and Severe Blood Disorders (Trinidad and Tobago) with children whose parents are members of the executive. The Society's first Gala Dinner Dance attracted 350 people to the Trinidad Hilton on 25 September this year.

Ia Bolton-Maggs, of Alder Hey Royal Liverpool Children's NHS Trust

bleeding which usually starts some hours later. In severe haemophilia A or B, where there is virtually no detectable factor VIII (in haemophilia A) or factor IX (in haemophilia B), individuals are liable to have bleeds into joints and muscles. Such individuals do not bleed spontaneously from other places (such as the nose or bowel) unless there is some other local cause. People with haemophilia DO NOT bleed abnormally from trivial cuts to the skin.

Haemophilia A, a deficiency of clotting factor VIII in the blood, occurs at a frequency of around 1 in 10,000 of the population, and the clinically similar disorder, Haemophilia B, a deficiency of clotting factor IX (Christmas disease - named after the first patient to be described with this disorder) occurs at a lower frequency of about 1 in 30,000.

HILIA B

abdomen or back. In patient D, the concentration of factor IX increased to 240 ng/ml from 71 ng/ml, maintaining a level of 220 ng/ml at present, expressing continuously for 6 months. The plasma activity of factor IX increased from 2.9% to 6.3%. The patient's bleeding tendency improved. In patient W, the concentration of factor IX increased from 130 ng/ml to 280 ng/ml, maintaining a level of 220 ng/ml with continuous expression for 5 months, but with an unstable increase. The two patients are being followed up. To date, no adverse side effect has been observed in either patient. We demonstrated that the whole procedure, including transferring the gene into autologous skin fibroblasts (mediated by retrovirus), covering cells with collagen and grafting under the skin, is safe, simple and feasible.

(Translated by Dr. Xing Chen with assistance from Dr. E.G.D. Tuddenham)

These are both inherited disorders, but in about a third of cases of haemophilia A no other affected males can be traced in the family. Haemophilia A and B are inherited as sex-linked recessive disorders.

Both haemophilia A and B vary in severity; some individuals are severely affected while others have mild disease. In general, the clinical pattern of the haemophilia can be predicted from the blood level of the affected factor, ie:

	Level of factor	clinical picture
Severe haemophilia A or B	<1 u/dl	Spontaneous bleeding into muscles and joints
Moderate haemophilia	2-5 u/dl	Less likely to have spontaneous bleeding Haemorrhage after Accidents surgery
and		
Mild haemophilia	5-50 u/dl	Bleed after surgery and accidents may never be diagnosed

That means that women can carry the abnormality and pass it on to both male and female children, but the disorder is usually expressed only in the male child.
The normal level of factors VIII and IX can be from 50 to about 150 u/dl.

It is important to know that haemophilia breeds true, in other words, if the type of haemophilia in your family is severe, then all affected individuals can be expected to be severe. On the other hand, if the haemophilia in your family is mild, then all affected individuals will be mild. This is important to consider when faced with the diagnosis, and perhaps in planning future children.

Another important fact to know is that carriers may have factor VIII or IX levels low enough to cause significant problems after injury or surgery. This is because although all women have two X chromosomes (see below), in all body cells one of these is randomly switched off. Because of this, on average a carrier will have a factor VIII level around 50 u/dl. However, some will be higher than average, because more defective X chromosomes are inactivated, but other women will have less than 50 u/dl, bringing them into the range of bleeding problems. About one third of carriers have levels of factor VIII below 50 u/dl. The practical consequence of this is, that if you are a carrier, you should know what your factor VIII or IX level is, in case you need special management to avoid excessive bleeding after surgery and dental extractions.

VIRAL TRANSMISSION OF HEPATITIS C AND HIV IN PARTNERS OF PEOPLE WITH HAEMOPHILIA

Heterosexual partners of haemophilia patients do not carry an exaggerated risk of HCV infection.

This, the principal finding of a study carried out at the Royal Free Hospital, largely confirms what has been observed in earlier investigations

Dr Telfer revealed the findings of research designed to determine the risk of heterosexual transmission of HCV and HIV in partners of anti-HCV-positive haemophilia patients 'Quite a lot is known about the heterosexual spread of HIV infection,' he said, 'but not so much about the heterosexual spread of HCV.

Although most studies suggest a low level of risk, a recent US study on the incidence of non-A, non-B hepatitis showed that in six per cent of new cases the principal risk factor was exposure to a sexual partner.

The Royal Free team tested the partners of 53 HIV-seropositive haemophilia patients, and the partners of 60 HCV-seropositive patients. Since 1983, couples attending the Royal Free have been advised to use barrier methods of contraception to reduce the risk of viral transmission. But median exposure to the virus was found to be 10.6 years in the case of the HIV patients and 17.3 years in HCV patients. 'This indicates,' said Dr Telfer, 'a long period when partners would have been exposed to the virus before we began to counsel them about the use of condoms.'

Four of the 53 partners (75 per cent) of HIV-seropositive haemophilia patients tested seropositive for HIV themselves. Only one partner of the 60 HCV-seropositive haemophilia patients (1.7 per cent) was found to be HCV seropositive.

Dr Telfer concluded that:

i) There is a low prevalence of HCV infection in partners of haemophilia patients.

ii) HIV is more prevalent than HCV in partners of patients with both infections - a finding that is in keeping with the fact that their partners have had prolonged exposure to hepatitis C and are mostly HIV seropositive.

He reported that the Royal Free is continuing to advise both their HCV- and HIV-seropositive patients to use barrier methods of contraception.

INFORMATION ABOUT DISABILITY WORKING ALLOWANCE

New information explaining the Disability Working Allowance has been published jointly by the Department of Social Security and the Benefits Agency.

A bright yellow booklet and purple leaflet together outline ways that people with an illness or disability who work more than 16 hours a week can top up their income.

The booklet says that people can qualify for the Disability Working Allowance whether they are working full time or part time, starting work for the first time, or returning to work after an illness.

The allowance is designed to help people with an illness or disability to gain new skills or experience or build up strength and stamina.

To qualify people must work 16 hours a week or more; have an illness or disability that puts them at a disadvantage in getting a job; must be getting a "qualifying" benefit; and not have more than £16,000 in savings.

The leaflet gives several examples of people who are currently receiving a Disability Working Allowance, while the 22 page booklet explains the qualifying conditions fully and includes a claim pack.

The booklet and leaflet are available at DSS and Benefits Agency offices, or people can call 0800-100-123 for an information pack. General information about benefit entitlement is available on the Benefit Enquiry Line on freephone 0800-882-200.

HAEMOPHILIA AND HEPATITIS C

from the British Society for Haematology annual scientific meeting

While the treatment of haemophilia patients has been revolutionised by the judicious use of clotting factor concentrates, inability in the early years to effect viral inactivation has had fatal consequences.

Thus, while spontaneous bleeding can now be treated effectively, and while surgery has become a safer procedure, as many as 60 per cent of patients who contract hepatitis as a result of blood transfusion go on to develop chronic hepatitis. Some 20 per cent of these patients develop cirrhosis over the course of a decade, often with resulting liver failure and hepatocellular carcinoma.

It was against this bleak background of chronic viral infection - notably HIV and non-A, non-B hepatitis - that Dr Paul Telfer, from the Royal Free Hospital, London, detailed a study looking at haemophilia patients treated with blood products.

"Potentially, this is a very large problem - but we really don't know very much about the natural history of non-A, non-B hepatitis; in particular, whether every patient with chronic hepatitis is likely to develop complications, and over what time period this may develop". In 1989, the virus responsible for most cases of post-transfusion hepatitis was identified as hepatitis C.

The Royal Free study looked at 1,220 patients with congenital coagulation factor deficiencies, of whom 269 were anti-HCV seropositive. The median age of the patients investigated was 32 (range seven to 82) Some 74 per cent had haemophilia A, 17 per cent haemophilia B six per cent von Willebrand's disease. Of the remaining three per cent, six patients were carriers, and one patient had Factor XI deficiency. Bleeding was classified as severe in 67 per cent and mild or moderate in 33 per cent.

HIV status was sero positive in 42 per cent and negative in 58 per cent. Eight patients (three per cent were hepatitis B surface antigen (HBsAg) positive Median duration of exposure to concentrates was 15.2 years (maximum 28 years, minimum three years).

Some 224 patients (83 per cent) had evidence of chronic hepatitis, eight (three per cent) had portal hypertension, nine (three per cent) had liver failure, while one (0.4 per cent) had hepatocellular carcinoma and 47 patients died.

The study revealed that patients older than the median, those who had been exposed to HCV for more than 15 years, and HIV seropositivity were all independent risk factors for chronic hepatitis. The severity of haemophilia, however, was not independently associated with an increased risk.

Dr Telfer concluded that

i) There are a large number of haemophilia patients who are anti-HCV seropositive as a result of blood product therapy

ii) Chronic hepatitis is present in 83 per cent of these patients - a higher percentage than has been reported in post-transfusion hepatitis C

iii) 68 per cent have been exposed to hepatitis C for at least a decade, and 12 per cent for more than two decades

"This is a disease that is still early in its evolution," he warned. Over the next two or three decades we may be seeing many more patients presenting with liver failure."

He urged that treatment should be directed towards such patients, and towards those who are also HIV seropositive.

JOINT CARE AND EXERCISES

Don't forget that the Society has a range of booklets on aspects of haemophilia care that are available to members and their families.

One of the most recent is Joint Care and Exercises. Prepared by the Haemophilia Chartered Physiotherapists Association, the booklet outlines simple ways of keeping your joints supple.

Joint Care and Exercises is available from the Haemophilia Society national office.